
Aus Klinik für Psychiatrie und Psychotherapie des Kindes- und Jugendalters
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DISSERTATION

**The Specific disorder of arithmetical skills.
Prevalence study in an urban population
sample and its clinico-neuropsychological
validation. Including a data comparison
with a rural population sample study.**

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INDEX

INDEX	3
Preface	6
1. Introduction	6
1.1. The importance of mathematics	6
1.2. A brief historical outline of mathematics	6
1.3. Definition of the Specific disorder of arithmetical skills	8
1.4. Research on the field of Acalculia and the Specific disorder of arithmetical skills	9
1.4.1. Research until 1960	9
1.4.2. Research from 1960 to present	12
2. Objectives of the present study	27
3. The screening process	28
3.1. Methods and design of the screening process	28
3.1.1. Description of the study population sample	28
3.1.2. Applied screening instruments	28
3.2. Screening Results	31
3.2.1. DRE 3 (Diagnostic calculation test for third grades)	31
3.2.2. DRT 3 (Diagnostic spelling test for third grades)	32
3.2.3. Teacher questionnaires	33
3.3. Discussion of the screening results	35
4. Clinical and neuropsychological validation of a suspected Specific disorder of arithmetical skills	37

4.1. Selection of probands with a suspected Specific disorder of arithmetical skills	37
4.2. Methods and design of the validation process	38
4.2.1. Clinico-neurological and psychopathological examination of the probands	38
4.2.2. Neuropsychological testing of the probands	38
4.3. Case summaries of the probands with a suspected Specific disorder of arithmetical skills	40
4.4. Discussion of the clinical and neuropsychological validation process	50
5. Data comparison with a study of the prevalence for the Specific disorder of arithmetical skills in a rural population sample	51
5.1. Outline of the Methodology of the rural population sample study	51
5.2. Results of the rural population sample study	51
5.3 Comparison of both studies	52
5.4. Discussion of the comparison	53
6. General discussion	55
6.1. The terminology of the Specific disorder of arithmetical skills	55
6.2. The validity of the diagnosis of a Specific disorder of arithmetical skills	56
6.3. Neuropsychological models	57
6.4. Diagnostic criteria for the Specific disorder of arithmetical skills	58
6.5. Conclusions	59
7. Summaries	60
7.1. Summary	60
7.2. Zusammenfassung	60

8. References	62
Lebenslauf	72
Danksagung	75
Eidesstattliche Erklärung	76

“Oh, don’t bother me,” said the Duchess;

“I could never abide figures!”

Lewis Carroll: ‘Alice in Wonderland’

Preface

In the following we shall refer to ‘mathematics’ not in its meaning as an academic science, but as the essential ability to carry out the basic arithmetical operations.

1. Introduction

1.1. The importance of mathematics

Power today is mainly power through and by the command of figures (Brüning, 1996). Mathematical power means having the experience and understanding to participate constructively in society (Romberg, 1993). Consequently, individuals who have deficits acquiring these important powers deserve attention and efforts to facilitate their access to mathematical knowledge. At the core of this help must be the understanding of their insufficiencies.

1.2. A brief historical outline of mathematics

The need for the institution of an universally applicable quantification first arose with the emergence of agriculture and animal breeding, because then the reaction to nature itself became insufficient. The interaction with and the planning of the environment originated the first mathematical terminology (Knaur, 1993).

This process continued with the development of trade and crafts. Fingers and toes obviously played a significant role in constituting numerical phraseology. In certain cultures, numerical systems were based on the values five (e.g. the language of the Khmer), ten (e.g. Arabic numbers) or twenty (e.g. language of the Mayas). Despite their different cultures, developmental stages and religions, the basic structures of the number systems of all these peoples are very similar. The first form of recording numbers was to carve a corresponding number of notches in a wooden stick. This evolved in the Roman and Egyptian cultures into a symbolic system with which any natural number could be symbolized. In these systems, a new symbol had to be used for every new decimal power. In other cultures, such as the Chinese, Sumeric and Indian, another method developed, in which the place of a figure determined its decimal value. India is also where the symbol and the figure for ‘Zero’ originated.

With the emergence of number systems the first arithmetical rules came to pass. But it was only the specialization of professions and slave labor in the Greek city-states that made scientific mathematics possible with well-known mathematic prodigies

such as Archimedes or Pythagoras. The first examples of mathematical demonstrations are known from that time.

Mathematics remained a science in the hands of a few for many centuries. M. Alchwarismi, an Arabic mathematician, published around 800 A.D. a compendium of mathematics for merchants called 'Algebra et Almucabala', creating the term 'algebra', Arabic for 'carrying over'. His own name later evolved to become 'algorithm'. The premise for a general distribution of mathematic science was created by the German A. Riese in the sixteenth century by his conception of principles for mental arithmetic. With the rise of industrialization and the introduction of mandatory schooling such an distribution of mathematical knowledge took place to an extensive scale. At the same time this process elucidates the ever increasing mathematical understanding in general: in the Middle Ages, multiplication was exclusively taught on universities.

In our time, electronic calculators and computers have come to dominate certain areas of mathematics. This also opened new areas of and possibilities for mathematical research. But it makes knowledge of mathematics far from unnecessary. As the United States Conference Board of the Mathematical Sciences pointed out: 'a strong mathematics education is at the basis of the nation's need for a competent workforce and an informed society' (CBMS, 1995). The scope of this statement can be extended to include all developed, if not all nations (e.g. Boissiere, Knight & Sabot, 1985; Rivera-Batiz, 1992).

1.3. Definition of the Specific disorder of arithmetical skills

In the present manuscript we are using the term ‘Specific disorder of arithmetical skills’ as defined by the World Health Organization in its ‘International Classification of Diseases, 10th Edition. Classification of mental and behavioural disorders: clinical descriptions and diagnostic guidelines’ (ICD-10). In its chapter F81: ‘Specific developmental disorders of scholastic skills’ the ‘Specific disorder of arithmetical skills’ is coded as F81.2 and defined as follows:

‘This disorder involves a specific impairment in arithmetical skills, which is not solely explicable on the basis of general mental retardation or of grossly inadequate schooling. The deficit concerns mastery of basic computational skills of addition, subtraction, multiplication, and division (rather than of the more abstract mathematical skills involved in algebra, trigonometry, geometry, or calculus).’

The ICD-10 then goes on to lay out the diagnostic guidelines for the Specific disorder of arithmetical skills as follows: ‘The child’s arithmetical performance should be significantly below the level expected on the basis of his or her age, general intelligence, and school placement, and is best assessed by means of an individually administered, standardized test of arithmetic. Reading and spelling skills should be within the normal range expected for the child’s mental age, preferably as assessed on individually administered, appropriately standardized test. The difficulties in arithmetic should not be mainly due to grossly inadequate teaching or to the direct effects of defects of visual, hearing, or neurological function, and should not have been acquired as a result of any neurological, psychiatric or other disorder.’ The disorder is distinguished from the ‘Acquired arithmetical disorder’, or Acalculia, coded as R48.8 in the ICD-10. Latter diagnosis stands for a loss of previously present arithmetical skills as opposed to the failure to acquire them in the former (WHO, 1992).

Unless otherwise noted, we will use the term ‘Specific disorder of arithmetical skills’ in the following if the criteria above apply, even if the quoted authors have used another terminology. This is meant to facilitate readability and understanding. For a discussion of the terminology ‘Specific disorder of arithmetical skills’ see chapter 6.1.

The other major classification system of mental and behavioural disorders which is used chiefly in Northern America, the Diagnostic and Statistical Manual of Mental Disorders, Fourth Edition (DSM-IV), has a parallel diagnosis with quite similar diagnostic criteria. In the chapter ‘Learning disorders’ the ‘Mathematics disorder’ is coded as 315.1. The DSM-IV then lays out the diagnostic criteria for the condition: ‘The essential feature of Mathematics Disorder is mathematical ability (as measured by individually administered standardized tests of mathematical calculation or reasoning) that falls substantially below that expected for the individual’s chronological age, measured intelligence, and age-appropriate education (Criterion A). The disturbance in mathematics significantly interferes with academic achievement or with activities of daily living that require mathematical skills (Criterion B). If a sensory deficit is present, the difficulties in mathematical ability are in excess of those usually associated with it (Criterion C)’ (Association, 1994). The similarities in the definitions of both classification systems are evident, certainly in their content, but in principal passages even in their wording.

1.4. Research on the field of Acalculia and the Specific disorder of arithmetical skills

It is noted by many researchers, as well as both in the DSM IV (Association, 1994) and the ICD-10 (WHO, 1992) that there is a lack of research on that field of study. At the same time, other disorders, such as dyslexia, another learning disorder on the field of language acquirement, received considerably more attention of the scientific community.

As an illustration of that circumstance we conducted a data-search in 'Medline', a database containing information on medical publications. A search for the keyword 'dyscalculia', a term often used synonymously for the Specific disorder of arithmetical skills, in the data for the years 1966-1996 produced a list of 81 publications. An analogous search for 'dyslexia', a synonym for the Specific reading disorder (F81.1, ICD-10), in 'Medline'-data for the years 1966-1996 found 3273 publications containing that keyword.

The research focus on the field of learning disorders has not changed significantly in the last years, either. A 'Medline'-search found 1 publication containing the keyword 'dyscalculia' and 81 publications containing 'dyslexia' in the database for 1997. This means, that from 1966-1996 as many publications concerning dyscalculia have been published as 1997 on dyslexia alone.

1.4.1. Research until 1960

Alkmaeon, a student of Pythagoras, physician and philosopher, who probably also participated in bisections of the eye, claimed around 500 BC the brain to be the central organ of the senses. Galen went around 200 AD even further and assumed there to be a specialization inside the cerebral cortex as well as the white matter. In the third century Christianity became the official religion of the Roman Empire and Christian doctrine was to dominate much of science. Its most popular philosopher St. Augustine (354-430 AD) moved away from the corporeal view of thought and claimed the existence of an immortal soul that can be influenced by but is independent of the body.

This view endured 1400 years and the brain as a site of human thought only regained interest when Descartes in the eighteenth century localized the 'animal spirits', responsible for muscle motion, blood circulation, respiration, sensory impressions, appetites, passions and memory in the cerebrospinal fluid. He agreed with the Christian concept of the soul, but thought it had to interact with the animal spirits in some corporeal site. This he proposed to be the pineal gland since it was the only singular part of the brain.

By developing the phrenological theory, F.J. Gall moved in the beginning of the nineteenth century beyond Cartesian principles. He was convinced that the superior human intelligence is owed to the greater development of the human cerebral cortex and that there is a specialization of the cerebral cortex into 'organs'. Gall thought that one could draw conclusions about the different regions of the brain by examining the outside features of the skull. Together with J.C. Spurzheim they popularized this view as 'organoscopy' or 'cranioscopy' as they themselves called it. It later became known

as phrenology. A convexity of the skull was assumed to reflect a well-developed underlying cortical gyrus and vice versa with regard to a concavity (Hunt, 1993). By those comparisons Gall and Spurzheim tried to localize a number of cerebral functions. Because they thought mathematicians to have a protrusion in the temporal area of the skull, just behind and above the eye (see Fig. 1), they concluded that the calculation abilities are located in 'a convolution on the most lateral portion of the external, orbital surface of the anterior lobes' (cited in Levin, Goldstein, & Spiers, 1993). By the middle of the nineteenth century, the phrenological view had become so popular that there were for example 29 phrenological societies in Great Britain alone.

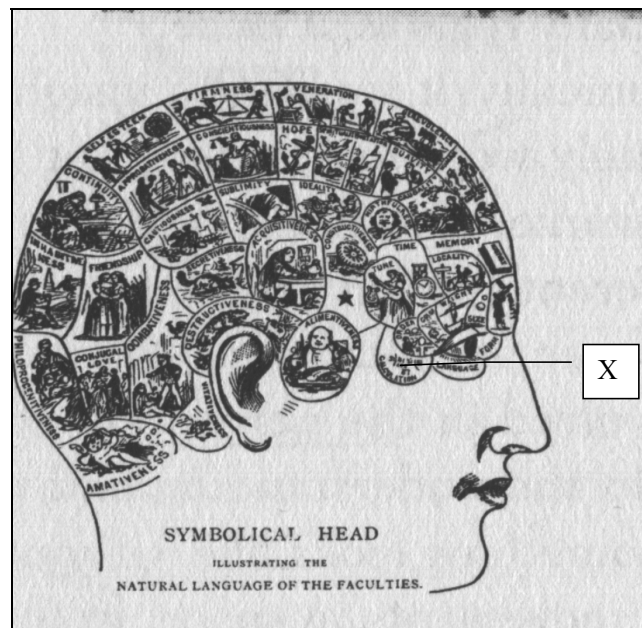


Fig.1 Contemporary illustration of phrenology. The proposed site for underlying calculation abilities is marked 'X' (from Hunt, 1993).

Although it is now known that the outer surface of the skull does not reflect its inner surface, let alone structures of the brain, it should be pointed out that almost 200 years later, a positron emission tomography study reported the involvement of the prefrontal cortex of the dominant hemisphere in visual calculation, a localization comparable to that of Gall and Spurzheim (Sakurai, Momose, Iwata, Sasaki, & Kanazawa, 1996).

To disprove the phrenologist theories which he thought lacked scientific methodology, the French physiologist P. Flourens developed around 1840 the experimental technique of ablation. In 1861 P. Broca and a few years later, in 1874, C. Wernicke introduced the clinico-anatomical correlations into neurophysiology and thus truly scientific methods were established in the field. These developments were greatly facilitated by new staining methods for neuronal tissue. Another important factor was the discovery of the X-rays by W.C. Röntgen in 1895 and their rapid utilization for imaging. It was then, that the first articles on arithmetical disorders and their site of localization in the brain were published.

Lewandowsky and Stadelmann described in 1908 a patient with deficits in addition, subtraction and the decimal system, who was found to have a hematoma localized at the left occipital side of the cortex (Lewandowsky & Stadelmann, 1908). They concluded that the calculation center of the brain is localized at that site. This meant two significant achievements: on one hand it was the first attempt to make a clinico-anatomical correlation in a disorder of arithmetic and Lewandowsky and Stadelmann were the first to describe arithmetical abilities independent from language abilities. They pointed out that in order to make a correct anatomic correlation, only cases of calculation disorders without aphasia should be considered. Seven years later, Poppelreuter described patients with shooting injuries to the head. He found arithmetical disorders in 12 of his patients. These also had a left- or double-sided hemianopsia. Concluding from the localization of the optic center, he considered the site of arithmetical abilities to be located in the cortex of the occipital lobes (Poppelreuter, 1915). In 1917 Sittig examined the calculation abilities of aphasic patients and proposed an influence of the left retrololandic area on number writing (Sittig, 1917). Peritz postulated 1918 the calculation center to be in the left angular gyrus (cited in Rüdiger, 1994).

The first systematic analysis of arithmetical disorders was conducted by Henschen, who coined the term *acalculia* ('Akalkulie'). He reviewed 305 cases with calculation disorders in the literature in addition to 67 cases of his own. Henschen stated that calculation abilities are to be considered separately from language abilities and their disorders. He concluded the existence of 'separate centers for letters (words) and figures (numbers)'. But he was also convinced that these centers would 'only with stimulation by adequate stimuli become active and conscious', thus postulating a cooperation of multiple regions of the nervous system (Henschen, 1919). Analogous to language production, Henschen assumed a motoric calculation center in the third convolution of the left frontal lobe and a sensory calculation center in the left angular gyrus (Henschen, 1925). But he also determined the need for further research and, in accordance with Lewandowsky and Stadelmann (1908), that only very circumscribed cases of *acalculia* should be analyzed in order to gain further information on the brain mechanisms involved in calculation (Henschen, 1925).

Soon after Henschen, Berger was the first to make a distinction between primary and secondary forms of calculation disorders. While primary calculation disorders develop independent from other cerebral disorders, secondary calculation disorders are those, that evolve as 'a consequence of damage or the loss of *other* cerebral abilities.' (Berger, 1926). Berger thought secondary calculation disorders to be more common. As causes for these impairments of arithmetical abilities he found attention deficits, memory and language disorders. In 18 of his own cases with the initial presentation of a calculation disorder he diagnosed after precise clinical examination only 3 cases of primary calculation disorder. In his publication he was the first to describe isolated deficits for certain arithmetical operations, namely division and multiplication. After an anatomical examination of his cases he concluded lesions of the temporal and the occipital lobe of the dominant hemisphere to be chiefly involved in cases of primary calculation disorders. Berger deduced a collaboration of these areas and possibly the frontal cortex of the dominant hemisphere in the acquisition of calculation abilities. A publication by Head, in which he proposed a terminology

linking a calculation disorder to each form of aphasia, is significant as the first publication on the matter by an English-speaking author (Head, 1926).

Gerstmann published an article in 1927 in which he described a symptom cluster devised of bilateral finger agnosia, right-left confusion, agraphia and acalculia (Gerstmann, 1927). This syndrome later came to be named after Gerstmann. No specific type of calculation disorder has been ascribed to the syndrome and the syndrome itself has been the subject of considerable controversy. Gerstmann later described the syndrome to be a consequence of lesions of angular gyrus of the dominant hemisphere (Gerstmann, 1940).

Leonhard described the role of spatial conceptions for arithmetical in a subgroup of 21 in 91 high-achieving individuals. He thought these people to have an internal representation of numbers and numerosity in a spatial fashion which they use for calculation procedures. The author makes a distinction between these 'figurative calculators' on one hand and the 'number-picture calculators' and 'number-word calculators' on the other. The 'figurative calculators' either calculate by counting (4 of 21 subjects) or by 'measuring' inside their internal numerical representation. In his paper, Leonhard pointed out that small multiplication tasks are solved by the use of rote verbal knowledge and not mental calculation (Leonhard, 1938). In 1948 Goldstein attempted to summarize the knowledge on calculation disorders in a book on language disturbances (Goldstein, 1948).

1.4.2. Research from 1960 to present

1.4.2.1. *Mental calculation in adults and acalculia*

1.4.2.1.1. R. Cohn

The neurologist R. Cohn was the first to attempt to develop a comprehensive model of calculation disorders. He chose multiplication tasks as the basis for his examinations (Cohn, 1961), and reasoned that five basic abilities are necessary for this mathematical operation: the recognition of numbers and operand, number ordering, a static memory for multiplication tables, a dynamic memory for carrying over results and addition. From case studies of dyscalculia he assumed 3 main causes of dyscalculia: disturbances in number ordering, memory problems and perseverations.

1.4.2.1.2. H. Hécaen and colleagues

Hécaen, Angelergues and Houllier strove for a more systematic model, formed on neurophysiological knowledge. Their model sought to segregate the calculation process in its components, then specify the errors resulting from malfunction of any of these components and finally ascribe these types of errors to lesions in particular regions of the brain (Hécaen, Angelergues, & Houillier, 1961). Hécaen et al. arrived at three different types of acalculia:

Type 1: Acalculia of the spatial type, in which the patients have problems to align digits correctly or maintain the decimal place of them or make errors such as

inversion, reversal or neglecting of numbers. The authors thought this form to be linked with a ratio of 12:1 to right-sided lesions.

Type 2: Acalculia resulting from alexia and agraphia for numbers, in which the patient is unable to read or write numbers. This form can occur independent of an inability to read or write linguistic material. Hécaen et al. thought this form to be correlated mostly with posterior, mainly left-sided cerebral lesions.

Type 3: Anarithmetria as the inability to calculate. This Hécaen et al. thought to be mostly associated with posterior, dominantly left-sided lesions.

1.4.2.1.3. Reaction time models of mental calculation

Groen and Parkman developed a model for calculation based on the measurements of reaction time for simple addition tasks. From their results with First-graders they deduced that the reaction time for a calculation task depends in a linear fashion on the complexity of the task itself (Groen & Parkman, 1972). After reaction time studies in adults they modified their model. They now proposed that the reaction time was composed of fact retrieval and counting. While results of certain calculation tasks are stored in long-term-memory and are simply retrieved in a constant time, the counting time for unknown tasks would vary depending on their problem size (Parkman, 1972). In addition to those results, Ashcraft and Battaglia found that the retrieval time for arithmetical facts stored in the long-term-memory also depends on their magnitude (Ashcraft & Battaglia, 1978).

1.4.2.1.4. Imaging studies for mental calculation

Besides the clinico-anatomical correlations for acalculia, attempts were made to obtain functional imaging of the calculation process itself. Sokoloff et al. attempted such a task with the nitrous oxygen technique and could not find any changes of global cerebral blood flow or oxygen consumption in mental arithmetic compared to baseline (Sokoloff, Mangold, Wechsler, Kennedy, & Kety, 1955).

More telling data only came forward with the advancement of functional imaging techniques. Roland and Friberg used radionucleotide imaging. They measured an increased cerebral blood flow in the prefrontal, inferior frontal and angular cortices of both hemispheres when they confronted subjects with a subtraction task (Roland & Friberg, 1985). In a positron emission tomography study (PET), Sakurai et al. demonstrated an increased activation of the left prefrontal and the left posterior superior temporal gyrus in 9 subjects when presented with multiple calculation tasks (Sakurai et al., 1996). Dehaene et al. made a more differentiated attempt in their PET study. They found that both in multiplication and comparison tasks, the occipital cortices are activated bilaterally as well as the left precentral gyrus and the supplementary motor area. In addition, multiplication only increased activation in both inferior parietal gyri, the left fusiform and lingual gyri as well as the right cuneus when compared to baseline whereas comparison yielded increased activation in the right superior temporal, the right middle temporal as well as the right superior and

inferior frontal gyri. They deduced an at least partial distinction of networks for both mathematical tasks (Dehaene et al., 1996).

The most recently published functional imaging study was again carried out by Dehaene and his colleagues. They assessed functional magnetic resonance images (fMRI) during approximation and exact calculation tasks in three male and four female probands. The fMRI showed greater activation in the parietal lobes for approximation than exact calculation. Specifically, the inferior parietal lobe showed increased activation in areas previously shown to be involved in tasks such as visually guided hand and eye movements, mental rotation and attention orienting. In exact calculation however, there was greater activation of mostly the left inferior frontal lobe, an area shown to be involved in verbal association tasks. The authors reach the conclusion that different cerebral networks are used in both tasks. Since fMRI has a low temporal resolution there is also an alternative interpretation of the shown data, namely that the activation differences are due to a secondary stage of mathematical reasoning rather than the primary information processing. Dehaene et al. rule this interpretation out with the employment of data from event related potential (ERP) in the same tasks. ERP have a high temporal resolution and the authors were able to show significant differences corresponding to the fMRI data in these tasks during the first 400 ms of a trial, when the probands were only presented with the mathematical tasks but not yet the choice stimuli (exact calculation vs. addition) (Dehaene, Spelke, Pinel, Stanescu, & Tsivkin, 1999).

1.4.2.1.5. Neuropsychologic models

The increase of data on mental calculation created a demand for neurophysiologic models. These have the advantage of yielding testable hypothesis and thus allow for a more detailed research. It is important though, that these models are modified once certain aspects of them have been found not to be true.

McCloskey and his colleagues analyzed a number of cases of 'acquired dyscalculia', or Acalculia. They specified the model of Hécaen and colleagues in an attempt to make more adequate neurophysiologic predictions. McCloskey and colleagues divided the process of calculation into two major modules, the Number Processing System and the Calculation System (McCloskey, Caramazza, & Basili, 1985).

The Number Processing System, in their model responsible for the perception of the necessary information, is subdivided into comprehension and formation of numbers, both composed of a verbal and a digit component. The Calculation System, responsible for the processing of the information is subdivided into a module for recognizing the operand, another for access to basic arithmetical knowledge and a third for the operation itself.

McCloskey et al. claimed that with this model they could not only describe their cases of acalculia but also make predictions about future cases and eventually find anatomical correlates for the different components (McCloskey, Aliminosa, & Macaruso, 1991; McCloskey, 1992).

Campbell and Clark countered the model of McCloskey et al. with their own Encoding Complex Theory of calculation. They argued that McCloskey et al. had failed to produce anatomical correlates of their proposed models and that several of the deficits described by McCloskey et al. may also occur in probands without deficits in calculation. They thus advised that the model should be abandoned (Campbell & Clark, 1988). Campbell and Clark suggested that mental calculation is a collaborative process of associated networks that relate through inhibitory synaptic transmission (Campbell, 1990). Consequently the authors concluded that disorders of calculation seldom occur solitarily, but most commonly in connection with other disorders (Clark & Campbell, 1991).

A more specific model of neuronal networks was devised by Dehaene and Cohen. They reviewed the literature on acalculia and its anatomical correlates as well as their own cases of acalculia and proposed a 'Triple-code model' for calculation (Dehaene & Cohen, 1995). Their model postulates three main representations of numbers in the brain:

- 1) a visual arabic code located to the left and right inferior occipito-temporal areas of the brain. It represents numbers as strings of digits. The function of this representation is multidigit operation and the evaluation of numbers are even or uneven.
- 2) a magnitude code, located in the left and right parietal areas of the brain. Here, numbers are represented as distributions along an oriented number line. This serves to evaluate of the quantity and proximity of numbers as well as their comparison.
- 3) a verbal code, located in the perisylvian areas of the dominant hemisphere. It represents numbers as sequences of words. By this representation arithmetical rote memory is accessed and the plausibility of results can be controlled.

With the triple-code model, Dehaene and Cohen were not only able to fit in the results of many previously described cases but could also make adequate predictions about the forms of acalculia connected with lesions of specific areas of the brain. Their model of verbal code and its use in arithmetical rote memory retrieval, first postulated by Leonhard (Leonhard, 1938), would also explain the strong interference between multiplication and object naming tasks, as described by Campbell (Campbell, 1994) and why multilinguals subjects retrieve to the language in which they first learned arithmetic when confronted with simple calculation tasks (Shannon, 1984). Dehaene and Cohen showed in detailed case descriptions that there is evidence for asemantic routes for transcoding numerals, i.e. patients with impaired calculation abilities whose ability to read and write down arabic numbers is preserved (Dehaene & Cohen, 1997). This contradicts the model of McCloskey et al. which claims the existence of a central quantitative representation, independent of its form. They further argued, that the model of McCloskey et al. fails to explain the dissociation of the sparing of certain types of operations (e.g. subtraction) when others (e.g. addition and multiplication) are impaired. Additional dissension against unique internal numerical representation comes from a case study of McNeil and Warrington. Their patient was able to perform simple oral addition and subtraction but his ability for written addition was severely impaired (McNeil & Warrington, 1994).

Further evidence for the triple-code model comes from a study Dehaene et al. carried out in bilingual probands. Eight students, fluent in both Russian and English, were trained on a set of approximate and exact additions in either language. When these trained tasks were subsequently tested the probands performed significantly faster on the exact addition tasks in the language they were previously trained in, regardless of the language itself. For approximation tasks the performance was equivalent in the two languages. The probands were then presented with new tasks in a similar numerical magnitude. The authors found that the subjects performed faster only on the previously trained exact calculations. Dehaene et al. draw the conclusion that these data are further proof for at least two independent arithmetical representations. While exact arithmetic relies on a language based representation, approximation tasks and advanced mathematical understanding relies more on a language independent conceptualization (Dehaene et al., 1999).

1.4.2.2. The development of calculation abilities in children with a Specific disorder of arithmetical skills

The Specific disorder of arithmetical skills, first described by Cohn (Cohn, 1968), differs in at least two important aspects from Acalculia, summarized by Rourke and Conway (1997):

First, the tasks involved in executing learned arithmetical skills differ from those involved in learning arithmetic. While in the former, information is to a large extent retrieved, in the latter, processes such as maturation of concept formation and adaptive reasoning skills are involved.

Second, there is evidence that the tasks involved in learning arithmetic are associated with right-hemispherical processes. It is only after the successful learning that arithmetical facts can be retrieved and executed by left-hemispherical systems. As a consequence, in the clinical cases involving adults with acalculia there will be a dominance of left-sided lesions, whereas in children also right-hemispheric lesions or dysfunctions will interfere with arithmetical learning.

1.4.2.2.1. Basic mathematics skills in children

The facts about the mathematical development of children have recently been summarized in a comprehensive book (Geary, 1994). Here, only the facts most pertinent to our study are referred to.

There is now considerable evidence that human infants already possess an impressive number of basic skills necessary for mathematics. Wynn demonstrated that infants can most likely even perform simple calculations. She showed a small number of objects to 32 infants with a mean age of 5 months. Then a screen was raised and the babies saw a hand either adding or taking one object away. After that, the screen was dropped, with either a correct or an incorrect number of objects on display. Wynn found that the infants looked significantly longer at the incorrect display, looking time being now a standard procedure in the testing of infant cognition (Wynn, 1992). In the light of these results Bryant (Bryant, 1992) raised the

question, whether Piaget (Piaget, 1952) was right, when he claimed that infants do not understand the concept of calculation.

Starkey and his colleagues found that infants can recognize the number of a small set of objects and can tell whether there has been a change in number. They can make a correct connection between the number of acoustic stimuli (i.e. drumbeats) and the number of simultaneously displayed objects. Infants were even able to correlate the number of acoustic stimuli with the number of displayed objects when the rate and duration of the acoustic stimuli were varied randomly. This led Starkey et al. to assume that infants possess a sense of numerosity (Starkey, Spelke, & Gelman, 1990). Mix et al. were not able to fully reproduce the findings of Starkey et al. They could only find a significant correlation to the infants' looking time when the acoustic stimuli were constant in rate and duration. They cast a doubt on the thesis that infants are already able to genuinely represent the numerosity of a set of objects (Mix, Levine, & Huttenlocher, 1997). In a recent review article, Geary gives an overview of 'Potential Biologically Primary Mathematical Abilities', citing not only studies in humans, but also in mammals and other animal species as evidence. The author concludes that the ability of numerosity, ordinality, counting, and simple arithmetic are likely inherent mathematical abilities, as they can be found independent from cultures and even in nonhuman primates (Geary, 1995).

The fact that infants already possess certain mathematics skills, raises the question which factors contribute to these abilities. Gillis et al. found in a study of 264 reading-disabled and 182 matched control twin pairs a significant genetic influence on mathematics performance score, with the genetic factors predicting the mathematical performance even slightly stronger than environmental factors (Gillis, DeFries, & Fulker, 1992).

1.4.2.2.2. Gender differences in mathematical reasoning ability

Benbow and Stanley published data on the sex differences for mathematical reasoning ability. In a first publication, they found a significant majority of boys among the mathematically precocious, which even increases through the high school years (Benbow & Stanley, 1980). They then extended their research to include all levels of mathematical giftedness. The results, however, were similar. By the age of 13 years Benbow and Stanley found in 39820 high school students a highly significant sex difference in mathematical reasoning ability in favor of boys across all levels of capability. This difference was most pronounced at the high end of the distribution: here boys outnumbered girls by a ratio of 13:1, even though girls and boys were matched by intellectual ability, age, grade and voluntary participation. No such differences for verbal abilities were found (Benbow & Stanley, 1983).

The reasons for this difference remain unclear. In their sample, Benbow and Stanley could not find gender differences in formal training in mathematics. Neither did their data support the theory of divergent gender socialization playing a significant role (Benbow & Stanley, 1983). The authors suspect that greater spatial ability in males might play a role (Benbow & Stanley, 1980). Another contributing factor might be a specific 'mathematics anxiety'. Hembree found this to have a negative effect on mathematics performance and to be more pronounced in girls (Hembree, 1990).

Schwank studied the problem-solving algorithms in girls from an educational perspective. The author comes to the conclusion, that girls approach problems in a conceptual way when boys use a sequential manner. Thus boys find their solutions 'in dialogue with the material' whereas girls will try to solve the problem as a whole (Schwank, 1990). Educational techniques are usually poorly adapted to develop conceptual thinking. Von Aster suggests that consequently, if a girl has a problem with a mathematics task, she should not be encouraged to 'try again' but to 'think it through again.' (von Aster, 1999). A review of the findings regarding gender differences was conducted by Geary. The author suggests that the sex differences are limited to biologically secondary mathematical domains. Geary concludes from the literature that there might be two main reasons why the male mathematical performance is consistently found to be better: more elaborate neurocognitive systems that support spatial navigation as a result of sexual selection, and different social preferences and styles. These advantages lead to a positive feedback mechanism and further interest in mathematics, increasing the male advantage in certain mathematical domains (Geary, 1996).

1.4.2.2.3. Hemisphere specialization and arithmetical abilities

There is sufficient evidence that there is a specialization between the hemispheres of the brain in their contribution to certain cognitive performance tasks (Geschwind & Galaburda, 1987). There are also reports of distinct patterns in hemisphere specialization, such that left-handers might have a less pronounced lateralization than right-handers, whereas males have a more lateralized pattern of cerebral organization than females. Also, intellectually gifted adolescents were found to rely more on right-hemisphere functioning in basic information processing (O'Boyle, Gill, Benbow, & Alexander, 1994).

Annett and Manning investigated the relation between arithmetical ability, hand preference and hand skill. In a general population sample of school children they found arithmetical ability to be positively associated with left-handedness in both sexes (Annett & Manning, 1990). Accessory to these results are the data of Peters, who found strong right-handedness to be significantly associated with a lack of mathematical giftedness (Peters, 1991).

O'Boyle et al. observed an enhanced activation of the right hemisphere in the EEG of mathematically gifted, left-handed males compared to normal controls when confronted with a basic processing task (O'Boyle, Alexander, & Benbow, 1991). They later corroborated these findings, when they showed that the finger tapping rate of both hands was reduced in 24 male, left-handed, mathematically precocious subjects while confronted with a verbal task. In 16 controls of average ability the verbal task reduced the tapping rate of the right but not of the left hand. The authors suggest that enhanced right hemisphere involvement is the physiological correlate of mathematical precocity in males (O'Boyle et al., 1994).

1.4.2.2.4. Didactic models of learning mathematics

Piaget devised a theory on the 'Child's conception of number' (Piaget, 1952). That theory was evolved by Aebli into a more comprehensive didactic concept (Aebli, 1973). He emphasized the process of learning mental calculation as an advancing abstraction from reality in four steps. Other didactic models of mathematics learning are analogous (e.g. Grisseemann, 1996).

The first step is an action including real objects. Already there is an abstraction, but it can be compared with experience. (e.g. 'If I have five apples and take three away, how many are there left?')

The second step is a symbolic illustration of the arithmetical operation. The realistic representation is modified into a more abstract form. (e.g. 'If I erase three of the five circles on the blackboard, how many are there left?')

The third step is the transformation of symbols into numbers. These have the advantage of universal applicability. (e.g. 'How much is $5-3$?')

As the fourth and final step Aebli identifies the automatization of known results through repetition.

1.4.2.2.5. Definition and demographic data on the Specific disorder of arithmetical skills

Cohn described in 1968 a group of 12 children with deficits in mathematics skills and coined the term 'Developmental dyscalculia' for their condition (Cohn, 1968). Slade and Russell described four children with a Specific disorder of arithmetical skills. They maintained it to be a primarily cognitive deficit (Slade & Russell, 1971).

The first systematic assessment of the Specific disorder of arithmetical skills was undertaken by Kosci. He defined it as a 'structural disorder of mathematical abilities which has its origin in a genetic or congenital disorder of those parts of the brain that are the direct anatomico-physiological substrate of the maturation of the mathematical abilities adequate to age, without a simultaneous disorder of general mental functions.' Kosci studied a population of 375 Slovak fifth-graders and found 24 (6.4%) of them to have a Specific disorder of arithmetical skills (Kosci, 1974). Four years later, Spellacy and Peter laid foundation to the criterion of discrepancy between arithmetical and general achievement to define the disorder in a publication about a group of 14 children with a Specific disorder of arithmetical skills (Spellacy & Peter, 1978).

Badian attempted to identify the prevalence of learning disorders in a study of 1476 schoolchildren of grades 1 through 8. He defined poor achievement as a score below the 20th percentile of the Stanford Achievement Test. The author found low reading achievement in 2.2%, low mathematics achievement in 3.6% and low achievement in both reading and mathematics in 2.7% of his sample (Badian, 1983). In total he identified 6.3% of schoolchildren to have a below-average achievement in mathematics. Häußer examined 181 schoolchildren in a rural area and detected 12 children with a Specific disorder of arithmetical skills, equaling a prevalence rate

of 6.6% (Häußer, 1995). The most comprehensive investigation of the demographic features of the Specific disorder of arithmetical skills was conducted by Gross-Tsur et al. They tested 3029 (75%) of the fourth-graders of the city of Jerusalem. Children were diagnosed with a Specific disorder of arithmetical skills if their arithmetic achievement score was equal to or below the mean score of children being two grades younger. Gross-Tsur et al. thus found a prevalence of the Specific disorder of arithmetical skills of 6.5% (Gross-Tsur, Manor, & Shalev, 1996).

Klauer used a slightly different approach when examining a sample of 546 third graders. He defined the Specific disorder of arithmetical skills as a discrepancy of two standard deviations between actual mathematics achievement and expected mathematics achievements as predicted by performance in other academic areas. The expected performance was determined by a regression analysis. Klauer thus arrived at a prevalence rate of 4.4% for the Specific disorder of arithmetical skills. In his sample, the condition was more prevalent in girls (Klauer, 1992). Lewis et al. found a prevalence rate of 1.3% for a Specific disorder of arithmetical skills and a prevalence of 3.6% of schoolchildren with combined mathematics and reading underachievement (Lewis, Hitch, & Walker, 1994).

1.4.2.2.6. Problem solving characteristics of children with a Specific disorder of arithmetical skills

A number of publications has been concerned with the way children with a Specific disorder of arithmetical skills access arithmetic problems. Bull and Johnston examined the short-term memory, processing speed, sequencing ability and retrieval ability of seven-year-old children. They found that processing speed was the most reliable predictor for the arithmetical abilities of the children. The authors conclude that arithmetic difficulties in children are mostly due to their inability to automate facts (Bull & Johnston, 1997). A study by Barrouillet and his colleagues of seventh-graders with learning disabilities demonstrated that these children did not only make more errors and had problems with fact retrieval but that they also had difficulties with inhibiting the retrieval of associations irrelevant to the given task (Barrouillet, Fayol & Lathulière, 1997). Jordan and Montani observed that third graders with specific difficulties in mathematics did make more errors than controls under timed conditions. When there was no time limit for the resolution of the given tasks however, there were no significant differences between both groups, but the group with difficulties in mathematics did rely more than the control group on primitive back-up strategies such as finger-use or counting. A third group with general learning difficulties made more errors than controls under any conditions (Jordan & Montani, 1997). The use of back-up strategies, immature arithmetical concepts and a low rate of instant fact retrieval in children with underachievement in mathematics was also found by other researchers (Geary, 1990; Geary, Bow-Thomas, & Yao, 1992; Hitch & McAuley, 1991; Swanson, 1993).

A transversal study of students with a Specific disorder of arithmetical skills was conducted by Ostad. He examined students with a Specific disorder of arithmetical skills in grade 1 (n=32), grade 3 (n=33) and grade 5 (n=36) and compared them to corresponding numbers of students without difficulties in mathematics. The author was not only able to confirm the finding that students with a Specific disorder of

arithmetical skills relied more on a small variety of rather primary back-up strategies, but also that the use of these strategies did not change significantly from year to year (Ostad, 1997). Similar to those results is a longitudinal 3-year prospective follow-up study of children with a Specific disorder of arithmetical skills by Shalev et al. Their results indicate that in almost half of the affected children the Specific disorder of arithmetical skills persists independent of gender, socioeconomic status, and educational intervention. This persistence is most likely in children with severe arithmetic disabilities and those with a sibling who also suffers from a Specific disorder of arithmetical skills (Shalev, Manor, Auerbach, & Gross-Tsur, 1998).

1.4.2.2.7. Neuropsychological models of the Specific disorder of arithmetical skills

In his careful examination of the subject, Kosc also proposed six subtypes of the Specific disorder of arithmetical skills, four of which are closely resembling those of Hécaen et al. (Hécaen et al., 1961), described above. In addition, he also proposed two additional forms of the Specific disorder of arithmetical skills: the 'practognostic' form with the inability to manipulate objects for mathematical purposes and the 'ideognostic' form, standing for the inability to understand mathematical concepts per se (Kosc, 1974).

Rourke and his colleagues examined in a series of publications from 1978-1993 the Specific disorder of arithmetical skills from a neuropsychological perspective. They divided children with specific learning disabilities into two groups: children with deficiencies mainly in graphomotor and spelling performance (subtype A) and children with an impaired mechanical arithmetics and spelling performance but normal reading and mathematics comprehension (subtype R-S). They then went on to distinguish the two groups as follows:

The subtype R-S children are characterized by deficiencies in the rote aspects of psycholinguistic skills and pronounced deficiencies in the more complex semantic-acoustic aspects of psycholinguistic skills. These deficiencies are more pronounced in children of younger age inside the group. Older children of the group have outstanding deficiencies in the semantic-acoustic aspects of psycholinguistic skills. Rourke et al. thought the R-S-group children to have a higher performance IQ (pIQ) than verbal IQ (vIQ). Their performance on visual-spatial-organizational, psychomotor, tactile-perceptile and nonverbal learning tasks is normal. Rourke et al. thought these children to be well adapted socio-emotionally.

The subtype A children were characterized by deficient performances on visual-spatial-organizational, psychomotor, tactile-perceptual as well semantic-acoustic tasks of novel material. These children have profound problems on nonverbal learning tasks and show no benefit from informational feedback or continued experience. They have a higher vIQ than pIQ. Socio-emotionally they exhibit deficiencies in the adaptation to novelty, social competence, emotional stability and activity level.

Rourke et al. assumed the R-S-group to be an expression of right-hemispheric functional disorders and the group A, or Non-verbal-learning-disorder (NLD) children, as being impaired by left-hemispheric disorders. Both subtypes can show

arithmetical disabilities, but the type of arithmetical problems of the groups will be different: while the R-S group will make more mechanical arithmetical errors the subgroup A children will show more severe arithmetical errors as well as difficulties with the spatial organization of numbers. Therefore the focus of assessment and therapy for both groups has to be substantially different (Rourke, 1993).

Von Aster tested the hypotheses of Rourke and colleagues in a sample of 41 psychiatrically referred schoolchildren. 20 of those children had a specific and 21 a combined disorder of arithmetical skills. He found no IQ differences for 21 of those children. 6 of 9 boys but only 1 of 11 girls with a specific arithmetical disorder were diagnosed with a Gerstmann syndrome. Von Aster found the highest rate of neurological soft signs among boys with a combined disorder of scholastic skills and the lowest among girls with a specific arithmetical disorder. Internalizing disorders were most frequent in children without an IQ-discrepancy or a Gerstmann syndrome. Girls with specific arithmetical disorders manifested almost exclusively internalizing disorders and boys mostly externalizing disorders. In conclusion, von Aster is only partially able to support the findings of Rourke et al. and argues for a gender difference in learning disabilities (von Aster, 1994).

Similar data to those of von Aster were published by Shalev et al. in 1995. 25 children with a Specific disorder of arithmetical skills who showed evidence for either right or left-hemispheric dysfunctions were thoroughly assessed medically and with both an arithmetical and a psychological test battery. Children with an abnormal perinatal history, neurologic disorder or signs of bilateral dysfunction were not included. The authors were unable to identify patterns of arithmetical errors that were specific to left or right hemisphere dysfunction. They concluded that there is no specific neuropsychological profile for children with a Specific disorder of arithmetical skills, but that input from both hemispheres is necessary for the development of arithmetical skills. In contrast to the suggestions of Rourke et al., the authors found that left hemisphere dysfunction had a more severe impact on arithmetical abilities (Shalev, Manor, Amir, Wertman-Elad, & Gross-Tsur, 1995).

Von Aster proposed a neuropsychologic model of arithmetical learning based on the triple-code-model by Dehaene and Cohen (see chapter 1.4.2.1.5.) In his model, arithmetical facts first pass through a basal processing mechanism. If processed correctly, the facts then meet a modular structure for numerical representation, the triple-code-model as outlined by Dehaene and Cohen. Based on the findings in very young children (see chapter 1.4.2.2.1.), von Aster proposes the magnitude code to be a congenital ability. The verbal code is then developed in early childhood and the visual-arabic number representation in school (von Aster, 1999) (see Figure 2).

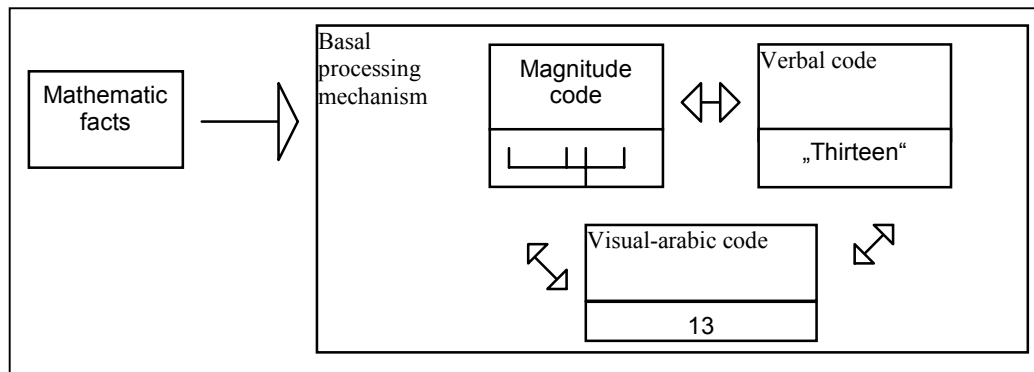


Figure 2: Triple code model for arithmetical learning

From that model, von Aster derives four different types of the Specific disorder of arithmetical skills:

In the first type, the basic processing mechanism is disturbed, resulting in global deficits in arithmetical abilities.

In the second type, the verbal representation of arithmetical abilities is deficient. Here especially the arithmetical abilities of counting and adding are impaired.

In the third type, the visual-arabic code is impaired. In this type of the Specific disorder of arithmetical skills especially reading, writing and comparison of written numbers are affected.

In the fourth type, due to a disturbed magnitude code, almost all arithmetical abilities are insufficient in spite of an intact basic processing capacity.

1.4.2.2.8. Socio-emotional characteristics of children with a Specific disorder of arithmetical skills

Most data on the socio-emotional characteristics of children with learning disabilities are not linked to a specific form of learning disorder. Waldie and Spreen examined a group of 65 learning disabled subjects who reported police contact at a mean age of 19 years. Six years later, they interviewed the same subjects again and then separated them into groups according to whether repeated police contact had occurred ($n=40$) or not ($n=25$). The authors found that of all the examined factors, only poor social judgement and impulsivity were reliable predictors, as subjects with high scores on these domains were much more likely to report repeated police contact (Waldie & Spreen, 1993). Huntington and Bender point out, that children with learning disorders attribute failure and success more to internal, personal factors and have a poorer self-concept (Huntington & Bender, 1993). Wright-Strawderman and Watson assessed 53 children with learning disabilities with the Children's Depression Inventory and found a prevalence of 19 children (35.8%) who scored in the depressed range (Wright-Strawderman & Watson, 1992). This is comparable to the rate of 26% found by Goldstein et al. (Goldstein, Paul, & Sanfilippo-Cohn, 1985) and the unpublished results of Wong (40%) and Chaskelson (29%) (cited in Wright-Strawderman & Watson, 1992). There are also indications that children with learning disorders are at an increased risk for suicide (Hayes & Sloat, 1988).

It seems that the characteristics, which constitute risk factors for the socio-emotional well-being of learning-disabled children, are prevalent in children and adolescents with a Specific disorder of arithmetical skills at a rate which at least equals that of all learning-disabled subjects. In addition to the results of Rourke et al. (see chapter 1.4.2.2.7.) other publications have been concerned with the socio-emotional characteristics of children with a Specific disorder of arithmetical skills. Johnson and Myklebust described a group of 14 children with a Specific disorder of arithmetical skills. They found them to be not well adapted socially, poor at estimating distance and time and lacking self-help skills (Johnson & Myklebust, 1967). Badian and Ghublikian examined 360 students and found that 16 boys with underachievement in mathematics relative to reading were rated significantly lower on a personal-social behavior skill by their teachers than two comparison groups. These ratings were very similar from both English and Mathematics teachers. They did not find significant personal-social characteristics for either girls with underachievement in mathematics or in any of the other groups (Badian & Ghublikian, 1983).

1.4.2.2.9. Comorbidity in children with a Specific disorder of arithmetical skills

Comorbidity is the co-occurrence of at least two different disorders in the same individual. The best-documented comorbidity for the Specific disorder of arithmetical skills is perhaps the co-occurrence of dyslexia. Ackerman et al. proposed that the majority of children with a reading disability evident early in childhood will go on to develop a deficiency in arithmetical skills in the higher grades (Ackerman, Anhalt, & Dykman, 1986). Badian found that almost half of the students with underachievement in mathematics have also a reading deficiency (Badian, 1983). Lewis found an almost threefold higher prevalence for a combined deficiency of scholastic skills than for underachievement in mathematics alone (Lewis et al., 1994) (see also chapter 1.4.2.2.5.) Gross-Tsur et al. found a prevalence of 17% for dyslexia in children with a Specific disorder of arithmetical skills (Gross-Tsur et al., 1996). The authors cited above hypothesize about the reasons for this comorbidity, but data for the analysis of the problem are scarce.

There is now considerable evidence of a genetic contribution to reading disabilities (DeFries, Fulker, & LaBuda, 1987) as well as mathematic performance (Gillis et al., 1992). Light and DeFries attempted to find the factors contributing to this comorbidity. They examined 259 twin pairs (149 monozygotic, 111 same-sex dizygotic) in which at least one twin had a history of reading problems and compared them to a sample of 134 monozygotic (MZ) and 93 same-sex dizygotic (DZ) twins without a history of learning problems. They found that 49% of the MZ and 32% of the DZ twins of reading disabled probands were mathematics disabled. Light and DeFries were able to show that approximately 26% of proband's reading deficits are due to genetic factors which also influence mathematics performance. The authors conclude that genetic and environmental factors contribute almost equally to the observed covariance in reading and mathematics scores (Light & DeFries, 1995).

Another disorder that is often observed in co-occurrence with the Specific disorder of arithmetical skills is the Attention Deficit Hyperactivity Disorder (ADHD). Shaywitz and Shaywitz noticed that children with ADHD often show problems in arithmetical achievement (Shaywitz & Shaywitz, 1984). In his aforementioned publication Badian

found that as many as 42% of children with low mathematics achievement showed evidence of attentional-sequential deficits (Badian, 1983). In a publication by Gross-Tsur et al. 26% of 143 children with a Specific disorder of arithmetical skills showed evidence for ADHD in their parent's or teacher's rating scales (Gross-Tsur et al., 1996). Stanford and Hynd found that learning disabled children and those with ADHD were rated equally by their teachers and parents on subjects such as daydreaming and inactivity. Only on closer questioning the observers perceived other forms of inattention and withdrawal in the two groups (Stanford & Hynd, 1994).

Evidence for further comorbid disorders is rare. But a publication by Shalev and Gross-Tsur indicates that this is more likely due to negligent observation than the absence of other comorbidities. The authors assessed seven third-graders with a Specific disorder of arithmetical skills who had not made significant academic progress despite their schooling in a special arithmetic class. They found neurological conditions with a direct influence on the children's cognitive abilities in all children. Four of these children had Attention Deficit Disorder without hyperactivity (ADD), one suffered petit mal-seizures, one ADHD and a developmental form of Gerstmann syndrome and one had severe dyslexia for numbers. Shalev and Gross-Tsur conclude that a thorough medical and neuropsychological assessment is necessary when the diagnosis of a Specific disorder of arithmetical skills is made (Shalev & Gross-Tsur, 1993), a view shared by others (i.e. O'Hare, Brown, & Aitken, 1991).

1.4.2.2.10. Etiological indications for the Specific disorder of arithmetical skills

Although the Specific disorder of arithmetical skills itself has not received as much attention as other learning disabilities (see chapter 1.4.) there is a considerable number of studies on the psychoeducational characteristics of other conditions indicating that a Specific disorder of arithmetical skills might be associated with these predicaments. In a recent review paper by Gross-Tsur et al. the authors conclude that: 'In fact, (the Specific disorder of arithmetical skills) is the most frequently encountered learning disability in children with epilepsy, fragile X carriers, Turner's syndrome and phenylketonuria.' (Gross-Tsur, Manor, & Shalev, 1993).

Other conditions should be added to that list. Klebanov et al. examined the school achievement of children with very low birthweight. They found those children with a birth weight below 1,000 grams to be most severely affected by risks such as grade failure and placement in special classes, even when they controlled for maternal education and neonatal stay. The only persistent academic difference of significance however, was in the mathematics score of the Woodcock-Johnson Achievement Battery. The authors controlled the group of normal birth-weight children to those of extremely low birth weight for intelligence. While the differences between both groups in reading achievement then abided, the extremely low birth-weight children still scored significantly lower than controls on arithmetical tasks (Klebanov, Brooks-Gunn, & McCormick, 1994). A study from Vohr and her colleagues suggests that mainly those low birth-weight infants might be affected that develop broncho-pulmonary dysplasia (BPD). While the full-scale IQ-scores of 15 BPD children were similar to those of weight-matched children the BPD children had a comparable reading but significantly lower arithmetic score on the Wechsler Intelligence Scale for Children - Revised edition (WISC-R) (Vohr et al., 1991).

Prenatal alcohol exposure also seems to affect the arithmetical abilities of children. Aronson and Hagberg made a follow-up of 24 children of alcoholic mothers. The authors discerned difficulties in mathematics, logical conclusion, visual perception, short-term memory and spatial relation abilities in these children (Aronson & Hagberg, 1998). Analogous to these results, Streissguth et al. found in a series of studies a strong correlation between maternal alcohol consumption during pregnancy on one hand and learning deficits as well as poorer spatial ability on the other. The level of impairment was dependent on the consumed amount of alcohol and the most prominent learning deficits were those in arithmetic (Streissguth et al., 1994a; Streissguth, Barr, Sampson, & Bookstein, 1994b; Streissguth, Bookstein, Sampson, & Barr, 1989). In a later study, the mathematics deficits of these individuals were examined more profoundly. Koperafrye, Dehaene and Streissguth observed particular difficulties in calculation and estimation relative to controls, but intact number reading and writing abilities (Koperafrye, Dehaene, & Streissguth, 1996).

Segalowitz and Brown questioned 616 high-school students about mild head injuries. They found that, contrary to a hospital-reported rate of 2-3%, there was a prevalence of head injuries of 31.2% among these students. They did not find an increased incidence of dyslexia or speech impairment in the group with head injuries, but they could show an association to depression, hyperactivity and mixed handedness. Although they did not assess academic achievement, the authors showed a correlation of head injuries and dislike of mathematics as a subject. This dislike was more pronounced the earlier the injury had occurred (Segalowitz & Brown, 1991). In the light of these data a case study by Levin et al. is interesting. They describe a 17-year-old boy who had sustained a right parietal skull fracture and right temporal hemorrhage at the age of 7 months. He showed a disproportionate impairment on arithmetical approximation and transcoding tasks, naming tasks as well as reading and spelling. In a fMRI study the authors were able to show that in this patient an early interhemispheric transfer of visuospatial skills, usually located in the right parietal area, to left parietal region had taken place (Levin et al., 1996).

2. Objectives of the present study

There are few demographic data on the Specific disorder of arithmetical skills. The current study was carried out to assess the prevalence of the condition in a general population sample of German schoolchildren using standardized academic achievement tests as diagnostic screening instruments.

Since the diagnostic criteria for the Specific disorder of arithmetical skills and their significance are widely discussed, we attempted in a second step of the study to validate the diagnosis of the Specific disorder of arithmetical skills from a neuropsychological and medical viewpoint. For the validation we assessed clinical data, imaging and neurophysiologic studies as well as a neuropsychological test battery.

Similar methods were used in another study of our research group of a general population sample of schoolchildren in a rural area. It was an additional goal of this study to compare these data with our data of a city sample.

Finally, we attempted to put our results into the larger context on the field of research of the Specific disorder of arithmetical skills.

3. The screening process

3.1. Methods and design of the screening process

3.1.1. Description of the study population sample

Four schools were randomly chosen in both of the two Berlin districts Mitte and Tiergarten from twelve basic-level schools. The two districts constitute the city center of Berlin. While Mitte is situated in the Eastern, Tiergarten is in the Western part of the inner city. With the choice of an equal number of schools in the East and the West we intended to control for any effect of different teaching methods or socialization in the formerly politically separate parts. Berlin, the German capital, is a major city with a population of 3.4 million people. 433,000 of its inhabitants are of foreign descent. The district Mitte has a population of 75,000 people, 9,500 of which are were not born in Germany. In the district Tiergarten 89,000 people are registered, with 24,000 inhabitants of foreign descent (Statistisches Landesamt, 1998).

In February of 1996, eight regular third-grade classes were included in the study. The sample included 182 third-graders, 100 male and 82 female. To minimize the effect of particular teachers on academic performance, all classes were from different schools. Special education schools were excluded. We obtained written permission for the study from the Office for School Affairs of the Berlin Senate, the school's principal and the teacher's and parent's of the included classes. We were not denied permissions for the study in any one student of the erstwhile chosen classes. The data were obtained in an anonymous fashion: the classes' teachers assigned a number to each proband. On the test sheets only those numbers were noted, and the teachers did not see the completed tests.

The age of the probands ranged from 8-11 years, with a mean of 8.73 years, the standard deviation (SD) being 0.69. There were 19.8% of probands ($n=36$), whose first language was not German. This likens the average population in Berlin, which has a quota of 23.6% for these pupils.

3.1.2. Applied screening instruments

The aim of the screening process was to identify probands with a likely diagnosis of a Specific disorder of arithmetical skills. Thus, the sensitivity of the method had relatively more importance than its specificity. The existing standardized German academic achievement tests for children are grade dependent and designed for use in either the last four months of the designated academic year or the first two months of the next one.

In spite of careful, long-term planning, starting in July of 1995, we obtained the written permission from the authorities cited above only in January 1996. The permission was limited to a period of two months, such as to minimize interference of the study with the academic schedule of the classes. The Berlin school year lasts until mid-July. Hence we were faced with the predicament that in the defined time it was either four months too late for the use of the standardized achievement tests for grade 2 or one month too early for the use of those for grade 3.

We eventually chose the test array of third-grade test for two reasons: first, because the temporal application criteria were met closer with those tests, and second, because our intent was not to assess the academic achievement of the probands per se, but to screen for achievement discrepancies with a high sensitivity.

3.1.2.1. Diagnostic calculation test for third grades (Diagnostischer Rechentest für 3.Klassen - DRE 3)

The DRE 3 can be used as an achievement test as well as a diagnostic test for arithmetical abilities and understanding. It is intended for use in the last four months of the third grade or at the beginning of the fourth grade. The test was standardized in 1971 with 2,975 probands. Its validity was ensured by using items that correspond to the academic curriculum for the third grade. In addition, the standardization sample was compared to the marks of the probands with good correlation. The test's reliability was measured at $r=0.91$ (Samstag, Sander, & Schmidt, 1992).

The DRE 3 contains 44 arithmetical problems, 40 of which are in arabic number format and 4 in story format. To maintain the proband's motivation the problems are first relatively simple and then gradually increase in their difficulty. The results for boys and girls differed significantly in the standardization sample. Accordingly, there are different test standards for both sexes. As the DRE 3 was designed as an achievement tests, its distribution has a right shift. Measured are correctly solved problems, with a mean of 28.79 for boys, SD 10.55 and a mean of 27.24 for girls, SD 11.08. The test can be evaluated by the quantity of solved problems. In addition, the character of errors can be assessed in a qualitative evaluation. There are two parallel test forms, such that adjoining probands can not copy results from each other. In order to avoid neglectful errors, the test is not timed. In all eight classes the probands were instructed and handed out the DRE 3 by the same person with the assistance of the teacher. The written instructions for the test's application were strictly followed. For further objectivity, congruity and comparability of the results, all classes were tested at similar morning hours. Probands who had completed the test were asked to quietly read or draw. After 70 minutes 179 of the probands (98.4 %) had completed the test. For three probands (1.6%) the test had to be finished after 75 minutes since the allotted time had run out.

3.1.2.2. Diagnostic spelling test for third grades (Diagnostischer Rechtschreibtest für 3.Klassen - DRT 3)

To distinguish low scores of arithmetical achievement representing a Specific disorder of arithmetical skills from those representing overall academic underachievement or low motivation, we assessed the spelling abilities with the DRT 3.

Analogous to the DRE 3 the former is an academic achievement test for third-graders. It is intended for use in the last four months of the third or the first three months of the fourth grade. A first version of the DRT 3 was standardized in 1965. The present version was standardized in 1982. In that later standardization the mean

error-score was almost 4 points greater than in the original edition. We used the third edition of the DRT 3, revised and using the 1982 standardization data of 3,690 probands.

The DRT 3 is constituted of 44 sentences with one word missing, respectively. The probands are handed out the test sheet. Then, the missing word in the first sentence is read by the examiner. Hereafter, the complete sentence is read aloud and finally the missing word is repeated. The test measures committed errors. As the DRT 3 is designed as an academic achievement test in accordance with the academic curriculum for the third grade, the score distribution is skewed to the left, with a mean error-score of 17.43, SD 10.08, found in 3,298 probands with German as their first language. For 392 probands, for whom German is not the first language, a mean error-score of 27.35, SD 9.18 was found. The test can be evaluated by quantity of committed errors and qualitatively, as evaluated by the character of errors (Müller, 1991). The test lasts between 25 and 30 minutes. In our study it was performed by the same person with assistance of the classes' teacher in all eight classes. The written instructions for the test's application were strictly followed. As with the DRE 3, the DRT 3 was carried out in all classes at a cognate time of the day.

3.1.2.3. Teacher questionnaire

Our study group developed a simple questionnaire for teachers. It was given to the teachers of each class. Since it is unlawful in Germany to submit student's marks to third parties, the teachers were asked to judge each proband's capacity in mathematics and spelling, respectively. A judgement of '1' meant the highest capacity in the academic field, and a '4' the lowest. With the questionnaire we strove to compare our data to the teacher's evaluation and to control for momentary difficulties of the probands. In one of the classes with 18 probands the teacher felt unable to fill out the questionnaire, as she had only recently begun to teach that class.

3.2. Screening Results

3.2.1. DRE 3 (Diagnostic calculation test for third grades)

The probands' scores in the arithmetical achievement test, separated by gender, are shown in figures 3 and 4.

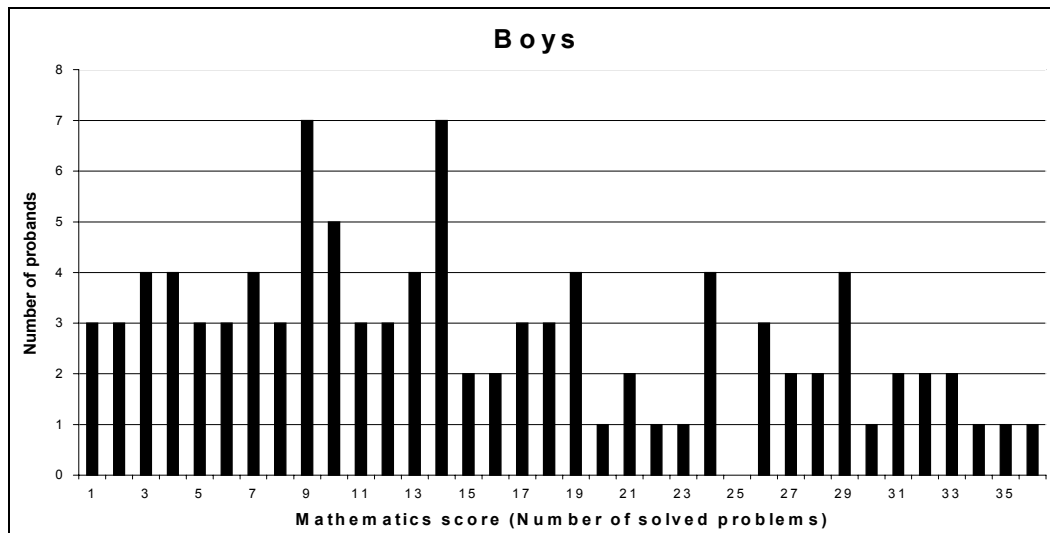


Fig.3: Arithmetical achievement scores for male probands

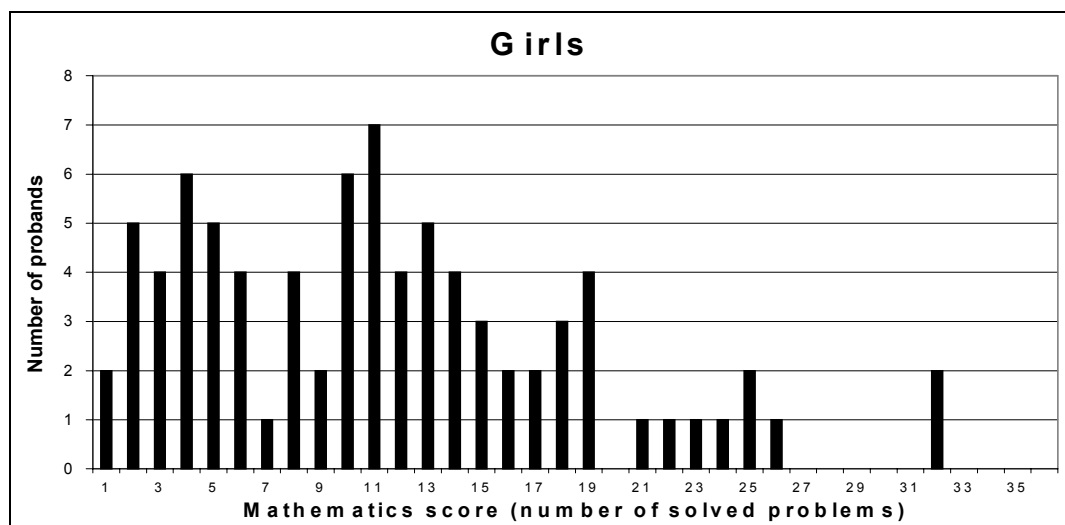


Fig.4: Arithmetical achievement scores for female probands

From these scores, the following distributions can be drawn (Table 1):

Tab.1: Average arithmetical achievement scores

	Total			Mitte district (East)			Tiergarten district (West)		
Sex	n	M	SD	n	M	SD	n	M	SD
male	100	14.6	10.14	47	14.57	10.51	53	14.62	9.2
female	82	10.38	7.24	44	10.34	6.92	38	10.42	7.67

(n: number of probands; M: mean DRE 3 score; SD: standard deviation)

3.2.2. DRT 3 (Diagnostic spelling test for third grades)

The probands' error-scores in the arithmetical achievement test, separated by German or foreign descent, are shown in figures 5 and 6.



Fig.5: Spelling raw scores for probands with German as their first language



Fig.6: Spelling raw scores for probands with German not as their first language

The resulting distributions of are shown in Table 2:

Tab.2: Average spelling raw scores

	Total			Mitte district (East)			Tiergarten district (West)		
First language	n	M	SD	n	M	SD	n	M	SD
German	146	26.31	10.03	84	24.32	10.13	62	29.01	9.31
Not German	36	32.69	6.82	7	31.00	10.92	29	33.10	5.63

n: number of probands; M: mean of committed DRT 3 errors; SD: standard deviation

3.2.3. Teacher questionnaires

3.2.3.1 Mathematics

Figure 7 shows the correlation of the score in the DRE 3 with the teacher's evaluation of the student's potential in mathematics.

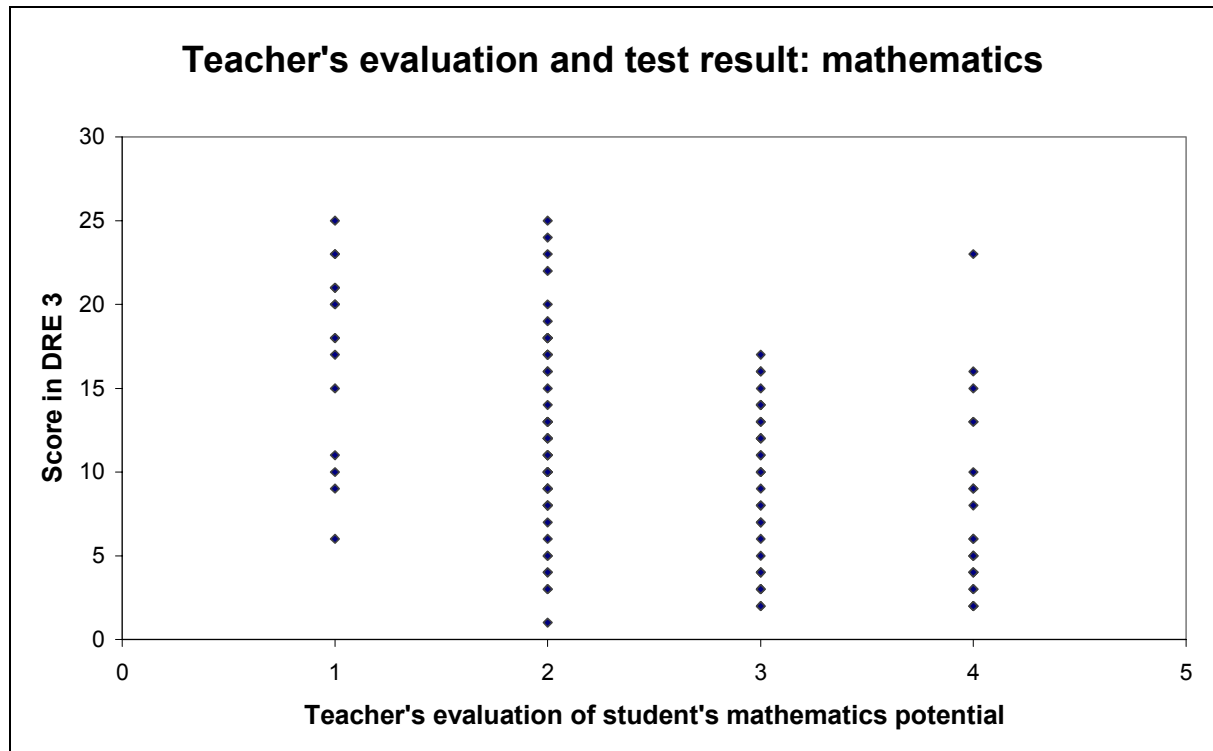


Fig.7: Teacher's evaluation of the mathematical potential of a student compared to this student's arithmetical achievement test result.

The figure shows some correlation between the teacher's evaluation of their student's mathematical ability and the corresponding test results in the DRE-3. However, test results in the lowest quartile of the tested probands can be found in the group of students judged by their teachers to be the most capable in mathematics (evaluation score 1) as mathematics scores twice as high and above the average can be found in the group with the lowest estimated potential for mathematics achievement (evaluation score 4). The correlation between the test results and the teacher's evaluation was calculated. Pearson's correlation coefficient was $p = -0.5788$.

3.2.3.2. Spelling

The correlation of the DRT 3 score with the teacher's evaluation of the students spelling potential is shown in Figure 8:

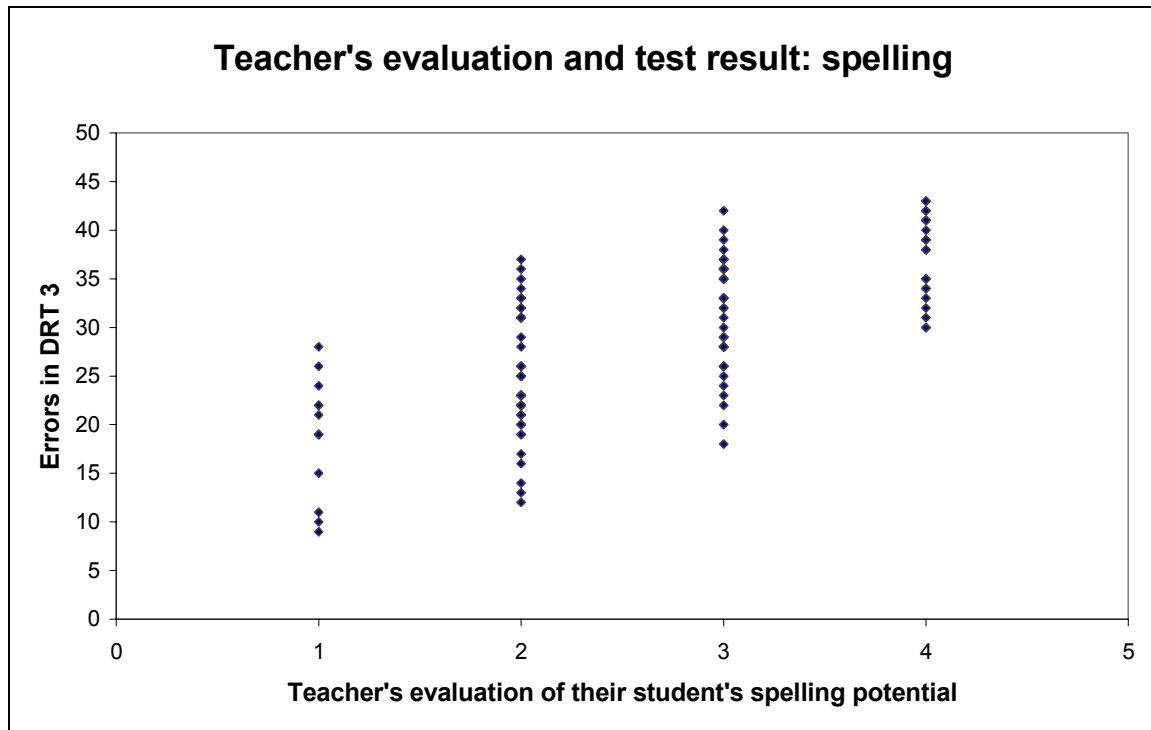


Fig.8: Teacher's evaluation of the spelling potential of a student compared to this student's spelling test result.

The figure shows some correlation between the teacher's evaluation of their student's mathematical ability and the corresponding test results in the DRT-3. However, there are test results just below the average of the tested probands in both the group of students judged by their teachers to be the most capable in spelling (evaluation score 1) as well as in the group with the lowest estimated potential for spelling achievement (evaluation score 4). The correlation between the test results and the teacher's evaluation was calculated. Pearson's correlation coefficient was found to be $p = 0.6039$.

3.3. Discussion of the screening results

In chapter 3.1.2. we explained our choice of screening instruments. Consequently, we expected the probands to score below the norms of the achievement tests. On the other hand, these tests are designed to assess classes' academic potential nationwide and they are oriented on the standardized academic curriculum for third grades. Only four to six weeks after our study the temporal criteria for the application of both the DRE 3 and the DRT 3 would have been met. Therefore we were surprised by the degree of low test achievement displayed by the our results.

The average value for boys in the DRE 3 is 28.79 points (SD 10.55). In our sample we found the mean score for boys to be 14.6 points (SD 10.14), a score 49.5% below that of the standardization sample. The results for the girls were analogous: in our study the girls reached a mean score of 10.38 (SD 7.24) in the DRE 3. This is 61.9% lower than the normal mean score of 27.24 points (SD 11.08) for girls. Although the probands reached significantly lower scores than the standardization sample, the standard deviations of the samples were similar. In addition, only 7 (7%) of the boys, and 2 (2.4%) of the girls scored above the 50th percentile of the norm.

Comparable to that were the results of the test for spelling achievement. Our probands with German as their first language made an average of 26.31 (SD 10.03) errors in the DRT 3. This means that compared to the normal values from the DRT 3 of an average 17.43 (SD 10.08) errors for that group, our probands committed 50.9% more errors. The probands with German not as their first language committed an average of 32.69 (SD 6.82) errors, 19.5% more than the normal value for this group of 27.35 (SD 9.18). The probands with German as their first language in our sample thus reached an average score that resembles the score for probands for whom German is not their first language in the standardization sample of 1982. As for the DRE 3 scores, the standard deviation of both groups were comparable to those of the norm. Only 23 (15%) of probands with German as their first language, and 6 (16%) of probands for whom German is not the first language, scored above the 50th percentile of the norm. Consequently, the results of the probands in our study for both tests seem to be distributed homogeneously and thus display their factual academic achievement.

As explained in chapter 3.1.2. we carefully avoided achievement tests that were relatively too easy since we intended to discern differences between arithmetical and spelling achievement. We were concerned that with the use of achievement tests for the second grade, the typical data shift towards high achievement scores in such tests would be too pronounced as to make out discrepancies in academic performance. Furthermore, the applied achievement tests have the advantage to test knowledge that is constantly accumulated over schooling time. To ensure good motivation of the probands, both test begin with more basic tasks and gradually increase in difficulty. However, our expectations were not met. The test results of all students were skewed towards the low end of achievement in both the arithmetical and the spelling domain. Since the obtained test scores had a homogenous distribution that was comparable to that of the standardization sample, our data seem to represent a realistic picture of the proband's academic abilities.

One explanation of our data would be a lack of motivation. But we could find no indication for that. Although the probands knew that the tests would not be graded, their application in the schools with the assistance of the probands' teachers granted an observably sufficient motivation. No proband terminated testing or displayed disruptive behaviour. Both tests start with easier tasks taught in earlier grades and only gradually increase in difficulty. To further avoid frustration, both tests include continuous positive verbal feedback regardless of results.

What then is responsible for the shift of data towards low achievement scores? The slightly early test administration alone does not seem to be a sufficient explanation. One contributing influence appear to be the relatively old standardization data. The DRT 3 had been last standardized 16 years, and the DRE 3 even 25 years prior to our application of the test. It is known that academic achievement scores in the Western world worsen over generations as IQ-scores increase, as it was shown of the USA in contrast to China by Geary et al. While the younger generation in both samples had higher IQ-scores, the arithmetic abilities of the American sixth-grade students were below those of American adults, in contrast to their Chinese peers (Geary, Hamson, Chen, Liu, Hoard, & Salthouse, 1997). Pfüller and Zerahn-Hartung showed that the orthographic achievement of German students significantly deteriorated in 27 years. They used a spelling test that was standardized in 1968. When they administered the same test in 1995, 48% of the probands scored below the 10th percentile compared to the probands that were tested 27 years earlier (Pfüller & Zerahn-Hartung, 1996). These data are quite analogous to our own findings.

4. Clinical and neuropsychological validation of a suspected Specific disorder of arithmetical skills

4.1. Selection of probands with a suspected Specific disorder of arithmetical skills

According to the diagnostic criteria for the Specific disorder of arithmetical skills quoted in chapter 1.3. a discrepancy of mathematics performance and other areas of achievement is required to make the diagnosis. Due to the shift of the screening data towards low achievement scores, described in chapter 3.3., it was problematic to identify probands with a significant discrepancy in their test results. The correlation between the teacher's ratings and their student's test results was equally too weak to be utilized as a diagnostic tool. Considering the low achievement scores, an over-evaluation of the probands' potential would be conceivable but under-evaluation of their abilities was just as common. We were not able to find a correlation between differences in test scores and corresponding differences in teacher's ratings.

Considering the homogenous distribution of our population's test results, we applied specific criteria in order to identify probands with a suspected Specific disorder of arithmetical skills. We selected those probands whose orthographic achievement was above the 50th percentile of our sample and whose mathematics achievement was below the 25th percentile of our sample. We thus arrived at the following criteria:

Tab. 3: Selection criteria for a suspected Specific disorder of arithmetical skills

First language	Sex	Score DRE 3	Errors in DRT 3
German	male	<6	<26
	female	<4	<26
Not German	male	<6	<32
	female	<4	<32

The criteria for a suspected Specific disorder of arithmetical skills were met by 12 probands, or 6.59% of the study population. Ten of these probands were girls. Eleven of the probands had German as their first language, six were from the Mitte district and six from the Tiergarten district.

The screening data were obtained in an anonymous fashion. In order to get in touch with the 12 selected probands we gave letters with the request for further participation to the teachers of the probands. We addressed the letters with the proband's code. The teachers then identified the probands and forwarded the letters to the families. The parents of five of these probands granted us permission for further investigation. Of these, three were girls and two boys all of them with German as their first language. For detailed case reports of the probands see below.

4.2. Methods and design of the validation process

4.2.1. Clinico-neurological and psychopathological examination of the probands

The five probands whose families agreed to further testing were given an appointment at the Clinic for Child and Adolescent Psychiatry and Psychotherapy of the Humboldt University Medical School, Charité Campus Mitte. A clinical history was taken and a thorough neurological examination with special attention to laterality preferences and other neurological questions was carried out.

If the parents gave their written permission, an electroencephalogram (EEG), visually evoked event-related potentials (VEP) and acoustically evoked event-related potentials (AEP) as well as cerebral magnetic resonance-imaging (cMRI) of the head were obtained.

4.2.2. Neuropsychological testing of the probands

4.2.2.1. *Attention and perseverance*

To evaluate attention and perseverance of the probands we used the Vienna Determination Device (VDD) equipped with Program R, a multiple-choice reaction test (Schuhfried, Berg, & Fischer, 1994). The proband's data were compared to a standardization sample of our clinic which included the data of 350 tested probands.

At the beginning and the end of the neuropsychological testing the proband works for two minutes with self-determined rapidity on the Vienna Determination Device. The task is to react swiftly and to discern between stimuli. The reaction time and the number of errors from both of the proband's test runs are compared and then evaluated in relation to the standardization sample.

4.2.2.2. *Laterality preference*

As shown in chapter 1.4.2.2.3., hemisphere specialization and hand laterality are crucial issues in the discussion of the Specific disorder of arithmetical skills. For this reason we tested the probands with the Motor Performance Series (MPS) (Schuhfried, 1994) using the standardization data of Sturm and Büssing (Sturm & Büssing, 1985). It analyses lateralization, dynamic coordination, accuracy and resting as well as diadochokinesia of the hands.

4.2.2.3. *Intelligence testing*

The intelligence of the probands was tested with two different methods. For an assessment of overall intelligence we applied a German version of the Hamburg-Wechsler Intelligence Scale for Children in the revised version of 1983 (WISC-R) (Tewes, 1984). We used an abbreviated form of it developed at Zurich University (WISC-R-A). It contains the subtests of Vocabulary (V), Arithmetic (A), Similarities (S), Digit span (DSp), Block design (BD), and Picture Arrangement (PA). The overall intelligence quotient (IQ) is then approximated with a regression formula (Bründler, 1989).

Von Aster et al. developed the 'Neuropsychological test battery for number processing and calculation in children (NUCALC)' to identify children with mathematical deficits. This specific test can be used from grades one through three and examines several basic skills that are necessary for arithmetic and also arithmetical abilities themselves. The examined basic skills are the counting of objects (CO), counting backwards (CB), estimating amounts (EA), estimating (EM) and judging magnitudes (JM), as well as the writing (NW), comparing numbers presented orally (NCO) or in written (NCW) and reading of numbers (NR). Arithmetical abilities are tested with mental addition (MA), mental subtraction (MS), and mathematical text problems (TP). The test was standardized for a German-speaking population sample. The subtests' scores can be translated into a quotient. A quotient between 85 and 115 represents an average ability. NUCALC provides the opportunity to identify and specify the profile of mathematical abilities in children with a Specific disorder of arithmetical skills. The instrument has been standardized for the ages 7-11 with a relatively small control sample. Aside from the age limitations and the small standardization sample, the divergent, sometimes considerably small, number of items per subtest (e.g. only 4 text problems), might be criticized and thus do not allow for a very detailed description of single deficits. Nevertheless, it represents a new quality in the description of arithmetic abilities (von Aster, Deloche, Gaillard, & Tièche, 1995).

4.3. Case summaries of the probands with a suspected Specific disorder of arithmetical skills

4.3.1.M.W.

Sex: female, age: 9 years, 10 months, first language: German.

Screening test results: errors in DRT 3: 9; score in DRE 3: 3.

Teacher's evaluation: German: 1; mathematics: 2.

4.3.1.1. Clinico-neurological findings

Family history: No known neuropsychiatric disorders in the family. No family history of learning problems.

Past medical history: Normal pregnancy and delivery. Birth at term without perinatal complications. Normal early childhood development. Developmental milestones met within normal temporal limits. *No significant medical or surgical history.*

Clinico-neurological examination: No abnormalities found on clinical examination. Normal, age-appropriate neurological status.

Psychological status: Fully oriented and alert. Friendly, cooperative girl. No pathological findings in psychological status.

EEG: Age-appropriately matured wave pattern. Right-hemispheric dominance over the occipito-parietal regions with normal function.

VEP and AEP: Physiologic latency periods with normal amplitude bilaterally.

cMRI: Permission denied.

4.3.1.2. Neuropsychological test results

Normal attention and perseverance as tested by the VDD.

Regular hand-lateralization towards right. Values in the average range for dynamic coordination, accuracy and resting. Diadochokinesia slightly below average.

Homogenous test profile in WISC-R-A with all subtests in the average range, lowest score on mathematical performance.

WISC-R-A-IQ=106. Subtest-scores (scores below average are in bold typeface)

V: 11 A: 9 S :10 DSp: 11 BD: 10 PA: 9.

Average performance in the test of number processing and calculation abilities. Only text-problem task performance slightly below average.

NUCALC Test-profile:

CO	CB	NW	MA	MS	NR	EA	NCO	EM	JM	TP	NCW
105	107	109	112	105	108	110	110	103	102	80	106.

In summary: no findings of pathological significance detected in the validation process. Mathematical performance was comparatively weak, however, the working diagnosis Specific disorder of arithmetical skills, as indicated by the results of the screening process, could not be validated. In the absence of other mathematical or cognitive difficulties, the below-average performance on the Text Problem in NUCALC is likely due to a diminished verbal memory capacity. No reason for the particularly low score in the DRE-3 could be detected.

4.3.2.Y.Z.

Sex: female, age: 8 years, 11 months, first language: German.

Screening test results: errors in DRT 3: 9; score in DRE 3: 4.

Teacher's evaluation: German: 1; mathematics: 2.

4.3.2.1. Clinico-neurological findings

Family history: No known neuropsychiatric disorders in the family. No family history of learning problems. Father speaks Turkish as first language.

Past medical history: Prematurely ruptured placental membranes in the fourth month of pregnancy, treated with tocolysis and mild sedation. Birth at term without perinatal complications. Normal early childhood development. Developmental milestones met within normal temporal limits. At the age of six weeks diagnosis and surgical correction of intestinal misplacement.

Clinico-neurological examination: No abnormalities found in clinico-neurological examination. Normal, age-appropriate neurological status.

Psychological status: Fully oriented and alert. Friendly, socially well-adapted girl. No pathological findings in psychological status.

EEG: Age-appropriately matured wave pattern. Hemispheric difference, especially over the occipito-parietal regions, pronounced under hyperventilation. Probable right-sided abnormality.

VEP and AEP: Physiologic latency periods with normal amplitude bilaterally.

cMRI: Hyperintensity of the occipital white matter as seen in myelinization irregularities (see figure 9). Otherwise normal, age-appropriate cMRI.

4.3.2.2. Neuropsychological test results

Normal attention and perseverance as tested by the VDD. Swiftness above average.

Regular hand-lateralization towards right. Values in the average range for dynamic coordination, accuracy and resting. Diadochokinesia slightly below average.

In WISC-R-A with most subtests in the high-average range. Vocabulary and logical thinking above average, visuo-constructive performance in the low-average range.

WISC-R-A-IQ=106. Subtest-scores (scores below average are in bold typeface) :

V: 15 A: 12 S: 15 DSp: 11 BD: 8 PA: 10.

No significant problems in number processing and calculation abilities.

NUCALC Test-profile (scores below average are in bold typeface):

CO	CB	NW	MA	MS	NR	EA	NCO	EM	JM	TP	NCW
105	107	117	112	99	113	112	116	104	116	119	110.

In summary: average to above-average results in most neuropsychologic tests. The working diagnosis Specific disorder of arithmetical skills indicated by the results of the screening process could be disproved in the validation process.

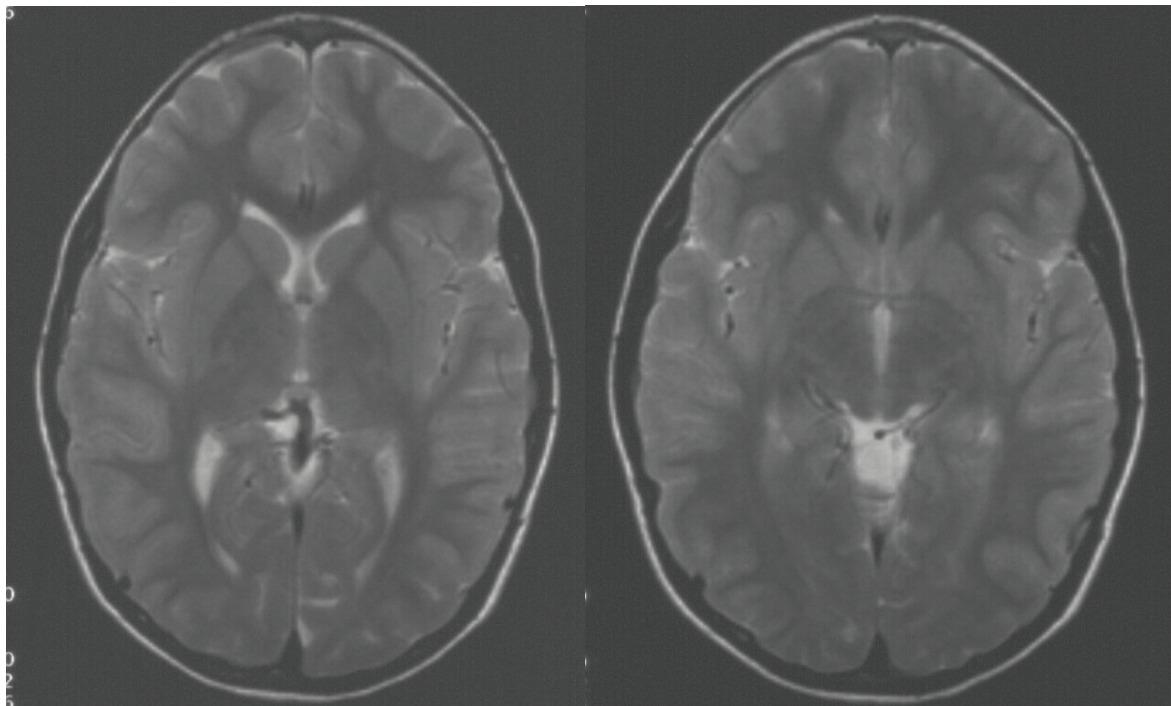


Fig. 9: cMRI of proband Y.Z. Note the poor differentiation between cortex and white matter in the occipital region. T2-weighted images.

4.3.3.S.P.

Sex: male, age: 9 years, 6 months, first language: German.

Screening test results: errors in DRT 3: 21; score in DRE 3: 6.

Teacher's evaluation: German: 2; mathematics: 2.

4.3.3.1. Clinico-neurological findings

Family history: No known neuropsychiatric disorders in the family. Both parents have a very good command of arithmetical tasks and had good grades in school mathematics. A 19 year-old half-sister of the proband from the mother's first marriage strongly disliked school mathematics, but had average grades. A 16 year-old half-brother from another father, born out of wedlock, is a mathematic prodigy and has an overall excellent academic performance.

Past medical history: Pregnancy originally was of twins, spontaneous abort of one fetus in the 3rd month of gestation. Mother was then admitted as a risk-pregnancy to the hospital and put under mild sedation. Birth induced at term without perinatal complications. Apgar (1/5/10 min): 8/9/10. Birthweight: 4200 g, length at birth: 52 cm. Normal early childhood development. Developmental milestones met within normal temporal limits. Cerebral commotion in 1995 with mild posttraumatic dysfunction shown on EEG. Gradual worsening of mathematics grades in school (Mark 1 in the second grade, mark 2 in the first semester of third grade, mark 3 in the second semester of third grade.) No past surgical history.

Clinical examination: Macrocephalic skull configuration, mild right-convex facial scoliosis. Inward-strabism of the left eye. Otherwise normal clinical examination.

Neurological examination: mild deficits in fine-motor coordination in upper and lower extremities on both sides. Normal reflex status. No focal signs. Normal, age-appropriate neurological status.

Psychological status: Fully oriented and alert. Slow reactions towards stimuli, absent-minded, daydreaming. Shy, contact-avoiding, internalizing. No other significant findings in psychological status.

EEG: Age-appropriately matured wave pattern without residues of the mild posttraumatic dysfunction of the previous EEG.

VEP: Physiologic latency periods with normal amplitude bilaterally.

AEP: Physiologic latency periods bilaterally of most waves. Wave 3 markedly delayed on the left side, indicating a dysfunction in the left pontine area.

cMRI: Normal, age-appropriate cMRI.

4.3.3.2. Neuropsychological test results

Normal attention and perseverance as tested by the VDD. Swiftiness above average.

Regular hand-lateralization towards right. Values in the average range for dynamic coordination. Accuracy and resting in the low average range. Diadochokinesia at the lowest testable limit.

In WISC-R-A most subtests in the high-average range. Vocabulary and visuo-constructive performance above average. Logical thinking, numeric short-term

memory and detection of visual contexts in the average range. Calculation ability in the low-average range.

WISC-R-A-IQ=110. Subtest-scores: (scores below average are in bold typeface):

V: 13 A: 9 S: 10 DSp: 11 BD: 12 PA 11.

Deficits in magnitude representation. No other significant problems in the test of number processing and calculation abilities.

NUCALC Test-profile (scores below average are in bold typeface):

CO	CB	NW	MA	MS	NR	EA	NCO	EM	JM	TP	NCW
70	107	109	111	89	108	110	101	103	102	107	76

In summary: mostly above-average results in most neuropsychologic tests. The arithmetical skills are distinctly weaker, most likely due to an impaired magnitude representation. However, the discrepancy between the mathematical performance and the overall performance is not sufficient to meet the ICD-10 criteria for a Specific disorder of arithmetical skills.

4.3.4.T.H.

Sex: female, age: 9 years, 2 months, first language: German.

Screening test results: errors in DRT 3: 20; score in DRE 3: 1.

Teacher's evaluation: German: 2; mathematics: 2.

4.3.4.1. Clinico-neurological findings

Family history: No known neuropsychiatric disorders in the family. Parents divorced. Mother works as a teacher, does not report any problems with mathematics. Mother's parents are both teachers for mathematics. No known family history of learning difficulties.

Past medical history: Diagnosis of maternal cholestasis in the 27th week of gestation, short hospital admission with ready clinical improvement. Birth at term. Length of labor: 4.5 hours, birthweight 3720 g, length at birth: 53 cm. Reflex-status diagnosed as pathological from 1 month of age. Delayed early childhood development: sitting upright from 10 months of age, first free steps with 16 months, first words with 18 months, first sentences at approximately 3 years. Diagnosis of a bilateral conduction hearing loss with 3 years and 8 months, treated with extracorporeal hearing aids with good success. Hearing loss stabile upon regular audiometric controls. No past surgical history.

Clinical examination: High palate, prognathia secondary to persistent thumb sucking. Inward-strabism of the right eye. Mild persistent dorsal positioning of the fingers ('Bajonettstellung'). On the inside of the left thigh: 3x1.5 cm depigmentation. Otherwise normal clinical examination.

Neurological examination: mild deficits in fine-motor coordination in upper and lower extremities on both sides. Normal reflex status of the upper extremities. Patellar reflexes lively bilaterally. Achilles tendon reflex more lively on right with mild after-discharge. Babinski's sign negative on the left side, mild dorsal flexion on the right. Mild tremor in Romberg's test. Mild gait insecurities, pronounced with blind gait. Marked misbalance in one leg-stand. No other neurological abnormalities.

Psychological status: Fully oriented and alert. Friendly and cooperative young girl. Repeated problems in understanding verbal instruction, constantly attempts to lip-read. Tendency to overplaying lacks in understanding. No other significant findings in psychological status.

EEG: Age-appropriately matured wave pattern. Suspected left-hemispheric lesion with regard to a overall reduction of amplitude and frequency on that side.

VEP: Physiologic latency periods with normal amplitude bilaterally.

AEP(assessed without hearing aid): Physiologic latency periods waves 3 through 5 bilaterally. Wave 1 markedly delayed bilaterally, indicating a peripheral impairment of perception.

cMRI: Two small hyperintensities, one located subependymal on the lateral side, right of the trigonum (see figure 10), the other on the medial, occipital aspect of the left posterior horn (see figure 11). Otherwise normal, age-appropriate cMRI.

4.3.4.2. Neuropsychological test results

Normal attention and perseverance as tested by the VDD.

Regular hand-lateralization towards right. Values in the average range for dynamic coordination. Accuracy and resting in the low average range.

Due to the hearing impairment especially verbal tasks had to be presented slowly, sometimes repeatedly. Upon persistent trouble with understanding, tasks were against standard procedure presented in a written form. The proband worked cooperative and diligent but markedly slow. Overall intelligence homogeneously notably below average, verbal and visual task performance approximately equal.

WISC-R-A-IQ=76. Subtest-scores (scores below average are in bold typeface):

V: 6 A: 6 S: 5 DS_p: 8 BD: 6 PA: 7.

Basic deficit in magnitude representation evident problems in the test of number processing and calculation abilities, with marked uncertainties and plural errors in more complex calculation tasks, such as operations with larger numbers, subtractions, and multi-step text problems.

NUCALC Test-profile (scores below average are in bold typeface):

CO	CB	NW	MA	MS	NR	EA	NCO	EM	JM	TP	NCW
72	107	112	112	81	113	36	102	72	87	73	75

In summary: Multiple findings indicative of a primarily left-hemispheric impaired cerebral function. Homogeneously decreased mental capacity with marked impairment of the arithmetical skills. The discrepancy between the mathematical performance and the overall performance is not sufficient to meet the ICD-10 criteria for a Specific disorder of arithmetical skills.

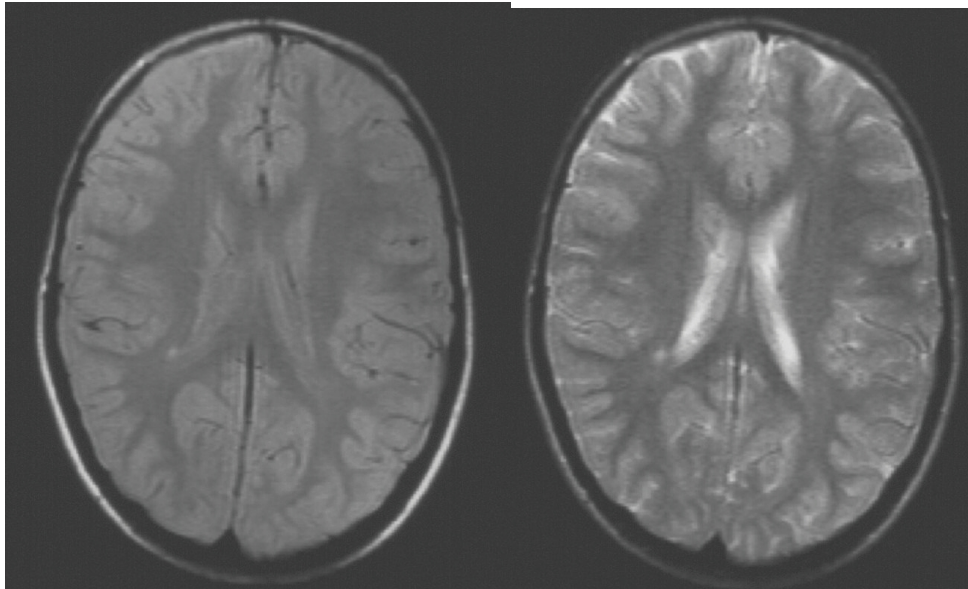


Fig. 10: cMRI of proband T.H. Note the hyperintensity located subependymal on the lateral side right of the trigonum. T1-weighted image on left, T2-weighted image on right side.

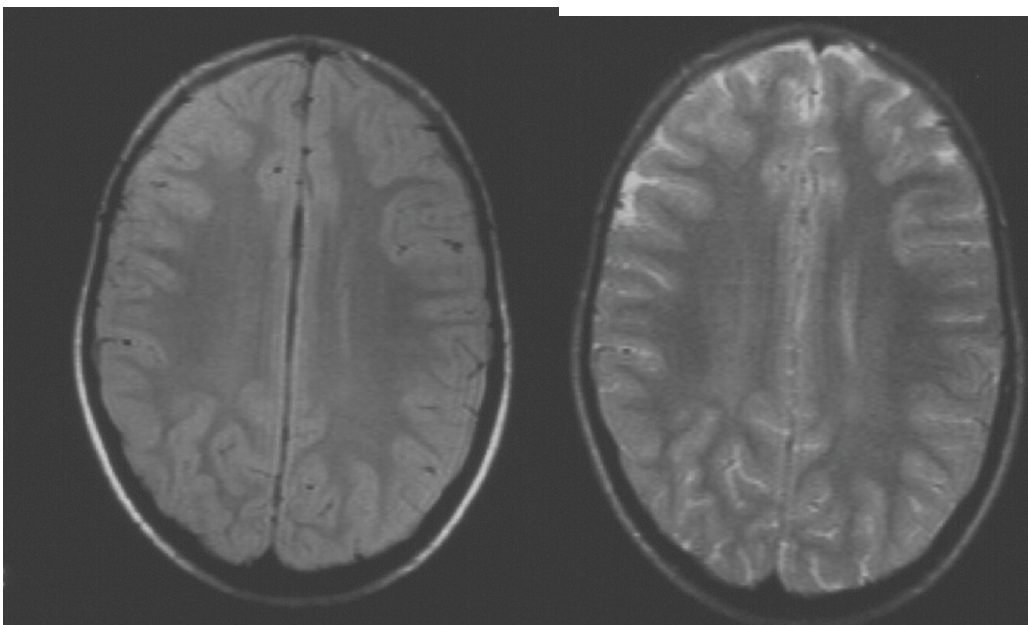


Fig. 11: cMRI of proband T.H. Note the hyperintensity located on the medial, occipital aspect of the left posterior horn. T1-weighted image on left, T2-weighted image on right side.

4.3.5.D.B.

Sex: male, age: 9 years, 8 months, first language: German.

Screening test results: errors in DRT 3: 22; score in DRE 3: 6.

Teacher's evaluation: German: 2; mathematics: 4.

4.3.5.1. Clinico-neurological findings

Family history: No known neuropsychiatric disorders in the family. Proband is child from the mother's first marriage, the father reportedly has little emotional understanding for him. Mother reports difficulties with mathematics throughout her time at school, always had to study extensively for average grades in mathematics. Has today command only of basic calculations. No other family history of learning problems.

Past medical history: Normal pregnancy and delivery. Birth at term without perinatal complications. Normal early childhood development. Developmental milestones met within normal temporal limits. Sleep irregularities since early childhood, needs two hours to fall asleep, talks, moves and sweats frequently at night-time, feels unrested in spite of adequate sleeping times. In the past six months frequent complaints of gastrointestinal discomfort and headaches. Highly motivated for school, increasing difficulties and deficits in mathematics notwithstanding. No past surgical history.

Clinical examination: Left-convex facial skoliosis. Otherwise normal clinical examination.

Neurological examination: mild deficits in fine-motor coordination in upper and lower extremities pronounced on the left side. Normal reflex status both extremities bilaterally. Otherwise age-appropriately normal neurological examination.

Psychological status: Fully oriented and alert. Friendly, but withdrawn boy. Shy, internalizing personality. No other significant findings in psychological status.

EEG: Age-appropriately matured wave pattern. Minor right-hemispheric lesion in the posterior temporal region.

VEP and AEP: Physiologic latency periods with normal amplitude bilaterally.

cMRI: Circumscribed leukomalacia in the white matter right of the trigonum with slight enlargement of the trigonum proximate to the lesion (see figures 12 and 13). Otherwise normal, age-appropriate cMRI.

4.3.5.2. Neuropsychological test results

Normal attention and perseverance as tested by the VDD. Proband worked on the VDD with markedly slowed speed.

Regular hand-lateralization towards right. Values in the average range for dynamic coordination. Accuracy and resting in the low average range. Diadochokinesia at the

lowest testable limit.

Very heterogeneous test profile in WISC-R-A, with an IQ in the low-average range, better performance on verbal tasks. Logical thinking is developed in the high-average range, the proband has an average vocabulary, but a calculation performance approximately 2 standard deviations below average. Visual performance was in the low-average range. Test profile consistent with a visuo-spatial processing deficit.

WISC-R-A-IQ=88. Subtest-scores (scores below average are in bold typeface):

V: 11 A: 8 S: 13 DSp: 9 BD: 6 PA: 6.

Basic deficit in magnitude representation and numerosity with marked uncertainties and plural errors on more complex calculation tasks in the test of number processing and calculation abilities, suggesting a relation to the weak spatial abilities. Proband is unable to check for plausibility even on relatively simple calculations (e.g. $12 + 6 = 16$). Frequent use of back-up strategies when faced with calculation tasks. Better performance on mathematical tasks with a strong verbal component.

NUCALC Test-profile (scores below average are in bold typeface):

CO	CB	NW	MA	MS	NR	EA	NCO	EM	JM	TP	NCW
104	107	109	64	81	43	86	110	103	110	95	106.

In summary: All findings indicate a indicative of a right-hemispherically impaired cerebral function. The discrepancy between the mathematical performance and the overall performance meets the ICD-10 criteria for a Specific disorder of arithmetical skills. In the light of the proband's internalizing personality it seems likely that his increasing somatic complaints have a strong psychosomatic foundation. Therapeutic intervention, both in the educational and medical realm are necessary to avoid a worsening of his situation.

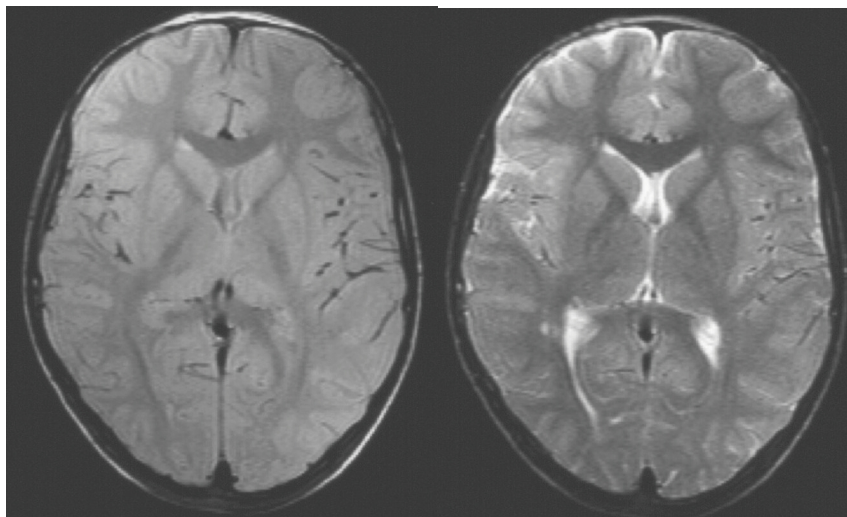


Fig.12.: cMRI of proband D.B. Note the lesion in the white matter, right of the trigonum with slight enlargement of the trigonum proximate to the lesion. The lesion is hypointense in the T1-weighted image (left) and hyperintense in the T2-weighted image (right).

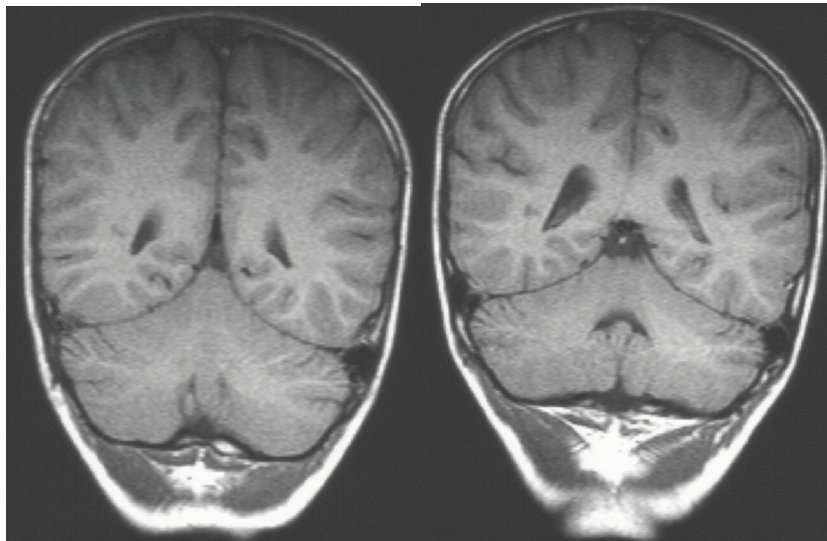


Fig.13.: cMRI of proband D.B., coronar slices of T1-weighed images. Note the slight enlargement of the trigonum, proximate to the lesion in the white matter, right of the trigonum.

4.4. Discussion of the clinical and neuropsychological validation process

We faced many problems with regard to the validation process, most of them laid out in chapter 4.1. A further problem was the low response rate, of 12 contacted probands only 5 (41.4 %) agreed to further testing. Even with repeated contact through the proband's teachers and the elucidation of the possible benefits of the examination process without any cost for the probands, we could not achieve a higher response. This is a common phenomenon and a well-known obstacle for research in Germany.

Considering these predicaments, our results were substantial. Only one of our probands (Y.Z.) had an altogether average performance on the test of number processing and calculation abilities (NUCALC). The other four probands performed below the average range on one or more of its subtests when compared to very recent standardization data from 1995.

In three probands (M.W., Y.Z., D.B.) the teachers rating corresponded well with the neuropsychological examination data. But in the two other probands (S.P., T.H.) we found remarkable discrepancies between the proband's results of their clinical and neuropsychological examination and the teacher's rating of their students of three years. Our data did not support the hypothesis of a previous publication of our group that the teachers are most likely to detect a Specific disorder of arithmetical skills in their students (Rüdiger, 1994).

To our knowledge there are no published data of imaging studies for children with a Specific disorder of arithmetical skills, although there has been a comprehensive debate about the hemispheric involvement in the disorder. We found two very similar lesions in two different probands with a mathematical performance markedly below average, pronounced in the domain of magnitude representation. Only further investigations will be able to show whether this finding is accidental or reproducible.

All tests and assessment methods utilized by us in the validation process yielded significant data, producing distinctive profiles of the probands. We dissent with the proposition that calculation difficulties should be exclusively considered as educational problems as stated by some authors (Grissemann, 1996). We agree with Shalev and Gross-Tsur that the working diagnosis of a Specific disorder of arithmetical skills should lead to a clinical and neuropsychological validation process (Shalev & Gross-Tsur, 1993). We would argue that such a validation is not only justified but should rather be obligatory before establishing the diagnosis.

5. Data comparison with a study of the prevalence for the Specific disorder of arithmetical skills in a rural population sample

As described in chapter 3.1.1., we examined an urban population sample in the present study. Preceding the present investigation, our study group had studied the prevalence of the Specific disorder of arithmetical skills in a rural population sample in 1995 (Häußer, 1995). The theoretical background and methodology were similar to those of the present study and are laid out in another publication by our group (Rüdiger, 1994). Therefore we attempted to compare the data of both studies.

5.1. Outline of the Methodology of the rural population sample study

Häußer carried out his study in the early fall of 1993 in the Dahme-Spreewald-district of the federal state of Brandenburg. Similar to the present study, laid out in chapter 3.1.2., the basic conception of Häußer for evaluating the prevalence was to assess the mathematics and language skills with standardized academic achievement tests. As defined in the ICD-10, an arithmetical performance significantly below the results of the language test led to the working diagnosis of a Specific disorder of arithmetical skills.

The study was conducted in eight third-grade-classes from August to September 1993, at the beginning of the academic year. The classes were chosen from four schools. A total of 181 probands underwent testing, 92 of which were boys and 89 girls. The Mathematics test for second grades (MT-2; Feller & Hugo, 1981) and the Diagnostic spelling test for second grades (DRT-2; Müller, 1983) were applied to the students, and their teachers were asked to evaluate their students performance in a questionnaire. The data were collected in an anonymous fashion.

An average or above-average performance is defined in the DRT-2 as a result above the 34th percentile of the standardization sample, and in the MT-2 as a performance above the 17th percentile of the standardization sample.

5.2. Results of the rural population sample study

The results in MT-2 and DRT-2 corresponded very well with those of the standardization samples of the tests, assessed in 1981 and 1983, respectively. The average value in the MT-2 standardization sample was for example a score of 52.47. Häußer found in his sample an average score of 52.4 in the MT-2. In order to make a working diagnosis for a suspected Specific disorder of arithmetical skills, the author consequently used the criteria of the MT-2 and DRT-2 to define discrepancies between academic skills. The criteria are shown in Table 4:

Tab. 4. Criteria for a likely learning disorder of the rural population sample.

Class of Learning disorder	Percentile MT-2	Percentile DRT-2
Specific disorder of arithmetical skills	<17	>34
Specific disorder of reading skills	>17	<26
Combined learning disorder	<17	<26

Twelve probands (6.6 %) met the criteria for a suspected Specific disorder of arithmetical skills. Of those nine (75 %) agreed to participate in a further clinical and neuropsychological validation process, which included a clinical and neurological examination, EEG, AEP, VEP and WISC-R-A as well as a test of hand-dominance. A Computer-assisted tomography (cCAT) of one proband and a common X-ray image of the skull of another proband were obtained. In the validation process only two of the probands were found to meet full diagnostic criteria for a Specific disorder of arithmetical skills of the ICD-10. Both of these probands showed left-sided functional deficits in the EEG and the clinical examination. In four probands the overall IQ was too low to fulfill the discrepancy criterion, and the arithmetical abilities of the remaining three probands were found to be within normal limits.

5.3 Comparison of both studies

There are many differences but also important similarities between the two studies of our research group, the present investigation of a city population sample and the one of a rural population sample by Häußer.

We conducted our study of the city population sample in winter, towards the end of the third academic year, whereas Häußer made his in early fall at the beginning of the academic year. Consequently, the present study used standardized academic achievement tests for the end of the third grade, while in the rural population study those for the second grade were applied. Another major difference is the fact, that in the former study our results of the standardized academic achievement test were significantly below those of the standardization sample, while the latter found good correlation of the data with the standardization sample. Table 5 shows a comparison of both population samples, when the standardization criteria for the achievement tests (see Table 4) are applied.

Tab.5: Comparison of the frequency of suspected Learning disorders in a city vs. rural population sample if the standardization norms of the screening instruments would have been applied.

Class of Learning disorder	Frequency (%)	
	Rural population	City population
Specific disorder of arithmetical skills	6.6	10.6
Specific disorder of reading skills	12.7	12.3
Combined learning disorder	3.3	48.6
No learning disorder	77.3	28.5

Another dissimilarity constituted the fact that there were no probands with another language than German as their first language in the rural population sample, while there were 36 (19.6 %) such probands in the city population sample. However, such probands were underrepresented in the group of probands with a suspected Specific disorder of arithmetical skills.

These important differences notwithstanding, there were also many similarities between the two investigations. Both studies were conducted with a practically equivalent number ($n=181$ vs. 182) of third-grade probands. No significant gender differences in academic achievement were found in either study and the results were homogeneously distributed. Most notably, in spite of different criteria and test instruments there was the same number of probands with a suspected Specific disorder of arithmetical skills, and the prevalence rates were practically identical in both studies (6.6 vs. 6.59%).

While we detected abnormalities with cMRI, Häußer found no abnormality in the two imaging studies he obtained. Since it is well-known that the cMRI is superior to other imaging techniques for information about the neuronal structure of the brain, we would argue for the cMRI as the imaging technique of choice when evaluating a Specific disorder of arithmetical skills in the absence of a clinically suspected gross anatomical abnormality.

5.4. Discussion of the comparison

The most conspicuous difference between both studies is probably the average performance of the rural population sample in the standardized academic achievement test in comparison to the below-average results of the city population sample in those tests. A number of reasons is responsible for that circumstance.

On one hand, the difference between both population samples is not surprising and it seems unlikely that only 28.5% of the probands of our city population sample do not have any form of a learning disorder. We applied the achievement test too early for reasons explained in chapter 3.1. It is therefore not unexpected that the test results were below the average of the standardization sample. Häußer, on the other hand, applied the MT-2 and the DRT-2 at the latest time permitted by the test criteria. It would thus have been conceivable that the performance of his population sample was above the standardization data. The data of both studies therefore confirm the findings of Pfüller and Zerahn-Hartung that the results in standardized achievement tests deteriorate over longer periods of time (Pfüller & Zerahn-Hartung, 1996). Furthermore the studies' results are in accordance to the German results of the Third International Mathematics and Science Study (TIMSS). The authors found that the German students reached the mean international results in mathematics on average 6 to 12 months later than their international peers (Baumert, Lehmann et al. 1997).

But the difference between the performance of both population samples is too salient, and the temporal difference in the test application does not seem to suffice as a solitary rationalization. As explained in chapter 3.3., our data of the city population sample do seem to be indicative of genuine lack in academic performance, whereas the performance results of the rural population sample merely could be better, but are in the average range. One difference between the populations was that the rural study population consisted exclusively of East German students as well as its teachers who were all trained in the former German Democratic Republic (GDR). In the teacher's education of the GDR teaching methods like pattern drill were emphasized and high standards of broad knowledge were implicated for all students. Those methods and goals had become unpopular in

West Germany during the nineteen-seventies and eighties, and there was a stronger focus on the individual achievement of each student. In the aforementioned TIMSS the authors found that a combination of both methods, as applied in many Asian countries, seems to produce the best results: to focus on the individual's achievement level in combination with ambitious educational intentions and consequent training. The authors infer that the individualization in Western European schooling has led to lowered expectations on the student's academic achievement and found a slight tendency toward better achievement in East German students (Baumert et al., 1997). At the same time these differences abide with time and none the probands of both studies had a conscious socialization in the GDR. The diminution of the East-Western difference was also noted in the TIMSS as this difference was more pronounced in older students who had more of their education in the former East Germany. In addition we found no difference in the city population sample between classes from the former West Berlin or East Berlin or between classes taught by teachers from either part of Germany. So differences in teaching methods between the formerly two countries fail to provide an explanation.

Another contributing factor to the stronger performance of the rural population is possibly the absence of students whose native language is not German in the sample of Häußner. In such classes, the teaching of academic skills in German is facilitated especially at the beginning of the schooling and thus could make for an educational advantage.

Other explanations are conceivable, such as a lower motivation of the inner-city population for education, a higher stimulus density in urban areas leading to an overflow of information or various environmental influences such as nutrition and air pollution. These factors were not controlled for in either of the two studies.

A common feature of both studies is that only one fifth of the probands (one in the present, two in Häußner's study) whose parents agreed to a clinical and neuropsychological validation process were found to meet the ICD-10 criteria for a Specific disorder of arithmetical skills, although the majority of the probands (four in the present, six in Häußner's study) was found to have significant and marked difficulties in their arithmetical performance.

The most important common feature of both studies is a prevalence rate of 6.6 % for a suspected Specific disorder of arithmetical skills according to the diagnostic guidelines of the ICD-10. This coincides with the findings of the publications by Kosci and Gross-Tsur et al. who both used a methodology and criteria similar to those of the study of Häußner and the present study for diagnosing a Specific disorder of arithmetical skills. The authors so arrived at a prevalence rate for the condition of 6.4 % and 6.5 %, respectively (Kosci, 1974; Gross-Tsur et al., 1996). This is a striking similarity, especially since these prevalence rates were found over a period of more than 20 years in three different countries with unrelated languages using different tests.

6. General discussion

6.1. The terminology of the Specific disorder of arithmetical skills

Many different names have been employed to label the inability of children to acquire basic arithmetical skills. In English publications the term 'developmental dyscalculia' (Cohn, 1968; Kosc, 1974; von Aster, 1994; Gross-Tsur, Manor, & Shalev, 1996), has been the original name for the disorder and is most frequently used, but also the singular 'dyscalculia' has been used synonymously (Spellacy & Peter, 1978; O'Hare, Brown, & Aitken, 1991). Other authors employ the names 'specific arithmetical difficulties' (Hitch & McAuley, 1991; Lewis, Hitch & Walker, 1994), 'specific mathematical difficulties' (Jordan & Montani, 1997), 'arithmetic learning disabilities' (Shekim & Dekirmenjian, 1978; Trembley, Caponigro, & Gaffney, 1980; Siegel & Feldman, 1983; Rourke, 1993), 'mathematic disabilities' (Garnett & Fleischner, 1987; Geary, 1993; Ginsburg, 1997) or 'arithmetic disorders' (Batchelor, 1989). Further authors, especially those embracing the neuropsychologic model of Rourke and colleagues, consider the inability to acquire arithmetic knowledge as cardinal symptom of the 'nonverbal learning disorder' (Rourke, 1989; Rourke, 1993; Brookshire, Butler, Ewing-Cobbs, & Fletcher, 1994;) or a symptom of the 'developmental right hemisphere syndrome' (Gross-Tsur, Shalev, Manor, *et al.*, 1995). In the German literature, the ICD-10 translation for the diagnosis is 'Rechenstörung' (calculation disorder) (Dilling, Mombour & Schmidt, 1993), but the terms 'Umschriebene Rechenschwäche' (circumscribed weakness in calculation) (von Aster & Göbel, 1990) and 'Dyskalkulie' (Grissemann, 1996) are also used.

This cacophony could be considered unimportant. Unfortunately it is of significance for several reasons. Without a common terminology it is markedly harder to make the condition better known to psychiatrists, psychologists, teachers and educators, let alone the general public, which leads to a low level of understanding for the children with the condition. This stands in striking contrast to dyslexia, where the affected children can often benefit from an extensive network of public health resources, special education and therapeutic interventions. This problem was already noted more than 20 years ago (Kosc, 1974). Since then, the importance of the problem has grown rather than diminished. The resources for special education are limited and families can only gain access to them on the basis of acknowledged diagnoses.

Secondly, each of the aforementioned terms are each defined slightly different by its authors. This makes it harder to compare the sparse data on the field and to find scientifically the most effective ways to treat the condition. Consequently, it would be highly desirable to have a common name and the same diagnostic criteria for the condition.

As described in chapter 1.3. we used in the present publication the name and the diagnostic criteria of the International classification system of the World Health Organization (ICD-10), as it came closest to the criteria of uniformity we desired. But, as we could show in chapter 4, and as it is argued by many of the leading researchers on the field (e.g. von Aster, 1994; Gross-Tsur *et al.*, 1996), at the core of the condition is an insufficiency to acquire arithmetic knowledge, caused by different factors. This can eventually lead to a 'Specific disorder of arithmetical skills'.

However, the loss of previously present arithmetical skills also leads to the clinical picture of a 'Specific disorder of arithmetical skills'. We therefore think that the term 'Developmental dyscalculia', chosen by the earliest authors and used by many of the leading contemporary authors of the field should be universally used, which beyond the fact that it is a historically and scientifically valid designation has the potential of wide international use.

6.2. The validity of the diagnosis of a Specific disorder of arithmetical skills

As mentioned in chapter 1.4. and noted by many authors, research on the field has been rather parsimonious for indeterminate reasons. Levine and his colleagues suspect, that since arithmetical knowledge is often equaled with intelligence, individuals with the disorder are simply considered less intelligent and consequently given less consideration. Furthermore, the authors argue, the overwhelming majority of people, including medical doctors, psychologists and teachers, did experience a level of mathematics, above which they did not fully comprehend the underlying concepts. This leads to the statement: 'I was never any good at mathematics, either.', underestimating the grave deficits of the affected children (Levine, Lindsay, & Reed, 1992).

In our sample we found more girls than boys with a suspected Specific disorder of arithmetical skills. As shown in chapter 1.4.2.2.2. there is much evidence that boys have an advantage over girls in mathematical reasoning ability. As opposed to dyslexia, it could be shown that the Specific disorder of arithmetical skills affects both genders at least equally (Gross-Tsur et al., 1996), if it is not more prevalent in girls (Klauer, 1992). In their socialization process girls are taught to rely more on quiet, internalizing problem-solving strategies, whereas boys are taught a more aggressive, externalizing approach. This could be an explanation why a learning disorder mainly affecting boys requires more attention in classrooms and subsequently in the medical and psychological communities.

The problems cited above have led some to question the existence of a Specific disorder of arithmetical skills altogether. Grisseman for example advises with regard to the problem 'to overcome the medical paradigm' (Grisseman, 1996).

We do not think that everybody who is slow in learning mathematics should be treated by specialists. The diagnostic criteria of the ICD-10 clearly demand that 'the arithmetical difficulties should not be mainly due to grossly inadequate teaching', an exclusion criterion firmly emphasized by us. There certainly is a significant number of students who fail to acquire adequate mathematical skills secondary to various deficits in their education, whether those are on an individual or a more general level. At the same time, we are convinced that the Specific disorder of arithmetical skills is a distinct diagnosis that falls into the medical domain. First, there is scientific prove that mathematical abilities and related disorders are genetically determined (see chapter 1.4.2.2.9.). Second, such different etiologies as very low birthweight, epilepsy, and early childhood alcohol exposure (among others) are known to cause the symptomatology (see chapter 1.4.2.2.10.). Thirdly, several studies in populations of divergent cultures and at different times have found quite comparable prevalence rates of the condition (see chapter 1.4.2.2.5.). And at last, the affected individuals

were found to display a distinct pattern of disturbance, when faced with arithmetical tasks (see chapter 1.4.2.2.6.). These facts are shown to be largely independent of educational systems or the individual's schooling.

These findings are supported by our own data. In chapter 4 we could show that there are children with a distinct inability to acquire mathematical skills. As other investigators (Kosc, 1974; Shalev & Gross-Tsur, 1993), we found accompanying medical conditions in some of our probands, could prove functional lesions and even found certain structural abnormalities with modern imaging techniques in others. We did not find a high correlation of the teachers' evaluations with their students' results on standardized academic achievement tests, or with the results of our thorough clinical and neuropsychological validation process.

We therefore conclude from our own findings, as laid out in chapter 4.4., as well as from the literature mentioned in chapter 1.4.2.2.9. that a careful clinico-neurological and neuropsychological evaluation process should be obligatory before the establishment of the diagnosis of a Specific disorder of arithmetical skills. We find this of particular importance, since the symptomatology has been shown to be persistent in longitudinal studies (Ostad, 1997; Shalev et al., 1998), and the socio-emotional well-being of the affected individuals is at risk (see chapter 1.4.2.2.5.). We do not mean to deprecate the intense and extensive need of these children for special education, which clearly falls into the educational realm. We only argue that at the beginning of the therapeutic process should stand an exact and thorough examination of the individual's resources, and that treatable medical conditions contributing to the diagnosis should be attended to.

Furthermore, as mentioned in chapter 6.1. we would find it desirable if the condition became better known among all professions concerned with the care of children, as a problems to acquire the 'intrinsically cumulative' (Levine et al., 1992) mathematical knowledge should be detected and appropriately approached as early as possible.

6.3. Neuropsychological models

Upon our clinical and neuropsychological examination of probands with a suspected Specific disorder of arithmetical skills, we were not able to support the neuropsychological model proposed by Rourke and colleagues, as outlined in chapter 1.4.2.2.7. (Rourke, 1993). Specifically, we found evidence for left-hemispheric as well as right-hemispheric functional deficits in the examined probands and were unable to find a persistent IQ pattern in our probands or similar deficits in psychomotor coordination, although there were probands with such a profile. Like von Aster (1994) and Shalev et al. (1995), we were not able to fit our data into the model of Rourke et al., or make adequate predictions about the clinical picture of the probands deficits with it.

Instead, our data corroborate the neuropsychological model of von Aster (1999) for the Specific disorder of arithmetical skills (see chapter 1.4.2.2.7. and Figure 2). While one proband (T.H.) displayed the first type of deficit according to this model (a deficient basic processing mechanism), another (M.W.) could be described with the second type (deficient verbal code), and two probands (S.P. and D.B.) suited the

fourth type (a disturbed magnitude representation.) The fact that none of the selected probands fitted the third type (deficient visual-arabic code) of von Aster's model might be due to the fact that these individuals not only have problems with reading and writing numbers, but also words. Our screening process would consequently either not have detected such probands or they are within the group of probands with a combined learning disorder.

Especially as the underlying triple-model of Dehaene et al. is increasingly substantiated by newer publications (see chapter 1.4.2.1.5.), and since our findings could be well predicted with the conception of von Aster, we endorse it as the currently most sufficient neuropsychological model.

6.4. Diagnostic criteria for the Specific disorder of arithmetical skills

The central diagnostic criterion for the diagnosis of a Specific disorder of arithmetical skills in the most common classification systems, the ICD-10 and the DSM-IV, is a significant discrepancy between the individual's general intelligence and his or her mathematical performance. From our findings we do not consider this to be a very good criterion. Although we did find distinct problems in solving arithmetical tasks in four of five examined probands, only one of them met the diagnostic criteria of the ICD-10.

One point of criticism with the criterion of discrepancy is that it is ill-defined. Most authors use our criteria, a performance of one standard deviation within the normal range in a standardized, individually administered intelligence test in contrast to a mathematical performance below in a standardized, individually administered academic achievement test. This procedure is advised in the ICD-10, but not obligatory. In other publications a mathematical achievement two grades below the actual grade of the proband has been used as a diagnostic criterion (Gross-Tsur et al., 1996), or all probands with an IQ below 90 were excluded (Lewis et al., 1994).

Even if the guidelines for the use of the standardized test would be specified, we still do think that this would not be the best possible way to diagnose the disorder. As other authors have noted, a majority of intelligence tests uses subtests strongly connected with mathematical abilities (Pfüller & Zerahn-Hartung, 1996). As a result, underachievers in mathematics will quite frequently have IQs below average, and can therefore not be diagnosed with a Specific disorder of arithmetical skills, such as our proband T.H. This was also noted by Kamphaus et al. who found that the standard method must produce an overproportionate number of children with a learning disability with above-average intelligence scores (Kamphaus, Frick, & Lahey, 1991). Other individuals do have a distinctly worse performance in arithmetical tasks compared to their overall intelligence (compare our probands M.W. and S.P.), but since their performance on mathematics tasks falls within the normal range of the standardization sample the diagnosis cannot be made either.

In a recent review, Rispen and van Yperen raise the same critical points against the discrepancy criteria for Specific Developmental Disorders of the common classification systems. The authors reach the conclusion, that the discrepancy should be abandoned and propose '...to develop diagnostic criteria that refer to

behavioral characteristics and underlying psychological and biological processes' of the Specific Developmental Disorders (Rispen & van Yperen, 1997). In another publication, Kulak analyzes the relatively larger body of research on the field of reading disorders and tries to draw conclusions for directions of research on the Specific disorder of arithmetical skills. She suggests that individuals with a severe form of either disorder do not only differ quantitatively in their acquisition of knowledge but in their quality. She further suggests a careful componential analysis of the skills involved in the acquisition of mathematical knowledge, as this has been a useful approach to reading disorders (Kulak, 1993).

Considering our data, we agree with the above authors that diagnostic criteria which are descriptions of qualitative symptoms are superior to a mere quantitative discrepancy criterion between ill-defined standards. Until these qualitative criteria are established, and when it is impractical to apply then, such as in the screening of larger groups of people, we favor the discrepancy approach of Kamphaus et al. They propose a regression analysis to define Learning disabilities, in which the individuals IQ performance is related with a regression approach to his or her performance in another test. The authors found that this approach generates a more homogeneous distribution of Specific learning disabilities in a study population (Kamphaus et al., 1991).

6.5. Conclusions

We conclude that there is a discrete group of individuals with severe problems in the acquisition of mathematical skills. These problems go beyond a quantitative range, but differ qualitatively from the ways unaffected individuals gain mathematical knowledge. These differences can be defined and predicted within a well-defined neuropsychological model supported by scientifically reliable data. We think that these qualitative differences should be best employed as diagnostic criteria rather than the discrepancy criteria used today. Since many, often treatable, conditions are known to lead to the disorder we hold that a thorough clinical and neuropsychological examination should be obligatory before establishing the diagnosis, especially since the affected individual's socio-emotional well-being is at risk. We believe that the term 'Developmental Dyscalculia' would be the adequate name for the condition.

7. Summaries

7.1. Summary

The Specific disorder of arithmetical skills is defined as an individual's inability to acquire arithmetical skills in spite of normal intelligence and adequate mathematical education. There is a considerable lack of data on the condition. The present study attempts to collect epidemiological as well as clinical and neuropsychological data.

In a first step, randomly chosen 182 third-graders of a city population were screened for a suspected Specific disorder of arithmetical skills, defined by a significantly worse mathematics compared to spelling achievement. We found the prevalence of the condition to be 6.59 % (n=12). In the second step we attempted to validate the suspected Specific disorder of arithmetical skills with a thorough clinico-neurological and neuropsychological test battery. Only five of the parents of the twelve probands agreed to further testing. Of those five, four showed below-average performances in at least one area of mathematical abilities. However, only one proband met the diagnostic criteria for the condition of the ICD-10.

The study compares our data with those of another study by our group of a rural population sample. Due to temporal differences and different screening instruments, the screening test results are quite dissimilar, yet the prevalence rate for the condition is equal in both studies.

Considering a review of the literature on the field as well as our own data the diagnosis of the Specific disorder of arithmetical skills is discussed. It is concluded that it has validity as a medical condition, but that is currently ill-defined. Diagnostic criteria employing qualitative aspects of the condition rather than quantitative discrepancies in standardized tests are proposed. An universal terminology of the condition is suggested and the need for further research emphasized.

7.2. Zusammenfassung

Die Rechenstörung ist definiert als Unfähigkeit, trotz normaler Intelligenz und angemessener Beschulung grundlegende Rechenfertigkeiten zu erwerben. Über dieses Störungsbild gibt es nur wenige wissenschaftliche Daten. Die vorliegende Studie hatte zum Ziel, epidemiologische sowie klinische und neuropsychologische Daten zur Rechenstörung zu erheben.

In einem ersten Abschnitt wurden 182 zufällig ausgewählte Drittklässler einer Großstadtpopulation mit standardisierten Leistungstests untersucht. Probanden mit einem erheblich schlechteren Ergebnis in Mathematik als in Rechtschreibung wurden als solche mit dem Verdacht auf eine Rechenstörung identifiziert. Wir fanden eine Prävalenz von 6,59 % (n=12). Der zweite Teil der Studie hatte eine umfassende klinisch-neurologische und neuropsychologische Diagnostik zur Überprüfung der Verdachtsdiagnose der Probanden zum Inhalt. Nur fünf der Eltern der zwölf identifizierten Probanden stimmten dieser weiterführenden Diagnostik zu. Von diesen fünf Probanden konnten bei vier umschriebene Störungen der Rechenfertigkeiten diagnostiziert werden. Jedoch nur einer erfüllte die diagnostischen Kriterien der ICD-10 für eine Rechenstörung.

Die erhobenen Daten werden verglichen mit denen einer anderen Studie der Arbeitsgruppe mit einer repräsentativen Stichprobe aus einem ländlichen Raum. Zwar finden sich aufgrund zeitlicher Unterschiede und unterschiedlicher zur Anwendung gebrachter Testinstrumente erhebliche Unterschiede in den Ergebnissen der standardisierten Leistungstests, die Prävalenz für Rechenstörungen ist jedoch in beiden Studien gleich.

Auf der Grundlage einer Übersicht bisher veröffentlichter Forschungsergebnisse und der erhobenen Daten wird die Diagnose der Rechenstörung diskutiert. Es wird geschlussfolgert, dass sie trotz teilweise unscharfer Definition als medizinische Entität valide ist. Der Vorzug der Verwendung von deskriptiv-qualitativen diagnostischen Kriterien an Stelle von quantitativen Differenzen in standardisierten Leistungstests wird betont, ebenso wie die Erfordernis einer international einheitlichen Terminologie. Die Notwendigkeit weiterer Forschung auf dem Gebiet wird unterstrichen.

8. References¹

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Lebenslauf

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1991 - 1993	Vorklinische Ausbildung an der Medizinischen Fakultät (Charité) der Humboldt-Universität zu Berlin
1993	Bestehen der klinischen Vorprüfung (Physikum)
1993 - 1994	1. und 2. klinisches Semester Humanmedizin an der Charité
1994	Absolvieren des 1. Abschnitts der Ärztlichen Prüfung; Note 4
1994 - 1995	3. und 4. klinisches Semester am Karolinska Institut, Stockholm
1995 - 1996	5. und 6. klinisches Semester an der Charité
1996	Absolvieren des 2. Abschnitts der Ärztlichen Prüfung; Note 2
September - Dezember 1996	Forschung; Tufts University Boston
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April - August 1997	Praktisches Jahr: Chirurgie

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	Pädiatrische Chirurgie: Floating Hospital for Children, Boston
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	Gastroenterologie: New England Medical Center, Boston
	Kardiologie: Oskar-Ziethen-Krankenhaus, Berlin
7.4.1998	3. Teil der Prüfung zum Staatsexamen:
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21.4.1998	Erhalt des Staatsexamenszeugnis Humanmedizin,
	Gesamtnote: 2,16 (gut)
8.5.1998	Erlaubnis zur Ausübung des ärztlichen Berufs als Arzt im Praktikum
10/1998 - 3/2000	Arzt im Praktikum an der Klinik und Poliklinik für Psychiatrie und Psychotherapie des Kindes- und Jugendalters der Charité Campus Mitte
01.04.2000	Approbation als Arzt
seit 01.04.2000	Wissenschaftlicher Mitarbeiter an der Klinik und Poliklinik für Psychiatrie und Psychotherapie des Kindes- und Jugendalters der Charité Campus Mitte
Forschung	
1996 - 1997	Collaborative Linkage Study for Autism bei Prof. Dr. Susan Folstein, Universitätsklinik für Kinderpsychiatrie, Tufts University, Boston; Forschungsprojekt zur Identifizierung der Gene, die zur Entstehung des Autismus beitragen; Studie fortlaufend
	Clinical-Neuroanatomical Comparison Study for Autism bei Prof. Dr. Margaret Bauman,

Harvard University, Boston; Erhebung standardisierter klinischer Daten und Vergleich neuroanatomischer Strukturen; Studie laufend

Self-Assessed Mini-Mental State Examination bei Prof. Dr. Marshal Folstein, Universitätsklinik für Psychiatrie, Tufts University, Boston; Pilotstudie zur Entwicklung eines Screeningtests zur Erhebung kognitiver Funktion

Veröffentlichungen

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Eidesstattliche Erklärung

Hiermit erkläre ich, dass die vorliegende Dissertationsschrift von mir selbst und ohne die Hilfe Dritter verfasst wurde, auch in Teilen keine Kopie darstellt und die benutzten Hilfsmittel sowie die Literatur vollständig angegeben sind.

Berlin, 12. Juni 2000

Jakob Hein